SAN FRANCISCO CHAPTER - LPA 1986 - 1990

Creator – Donna Hughes

Thanks a Million

By Percy Ross

DEAR MR. ROSS: I need a reliable used car. Let me explain. I'm 27 and do janitorial work which requires that I go from job to job. I use the bus as a means of transportation and sometimes must change buses two or three times just to get to one job. Even then, I can't get to many places where the better jobs are.

For many people this may not pose a problem. But I'm a dwarf and getting on the bus is a problem. The steps are so tall, that I must crawl to get onto them.

If I had a car, my income would increase and life would be so much easier. A friend has offered to make the special peddles for me, thus enabling me to drive.

Again, what I really need is a used car. If you can help, Mr. Ross, I'd be grateful. — Miss J.L., Seattle, Wash.

She needs a car to get better jobs

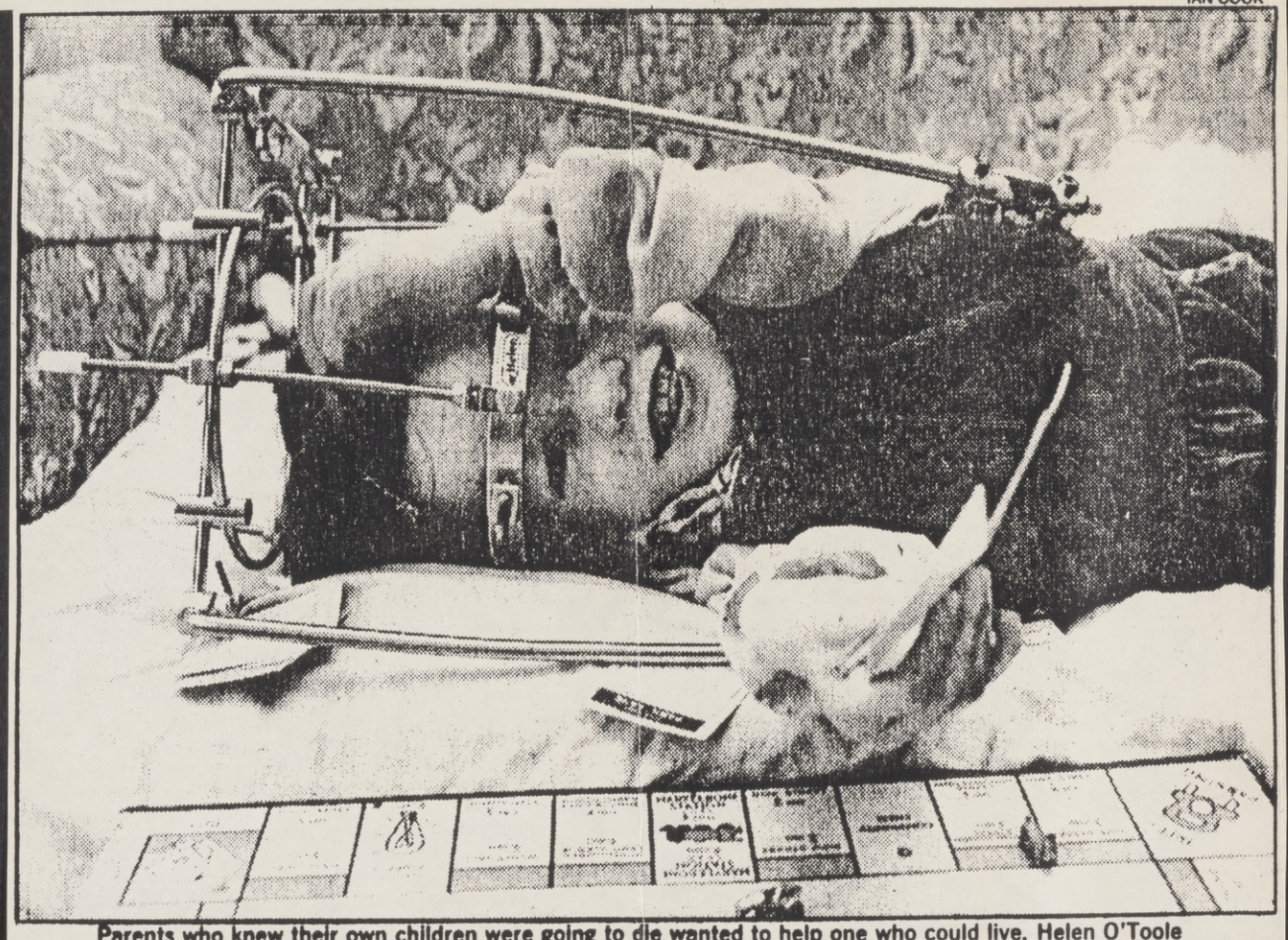
DEAR MISS L.: Throw away your bus schedule! From the reputable Lake City Auto Sales on Lake City Way NE, I've selected a dependable Pontiac Astro station wagon, which should be ideal for your needs.

Increasing your income is great . . . but this also should provide you with a new found sense of independence. Good luck!

THE SUNDAY TIMES 16 FEBRUARY 1986

Battle of the little people

IAN COOK



Parents who knew their own children were going to die wanted to help one who could live, Helen O'Toole

DENISE WINN tells the story of one child's rescue from an incurable disease

"MUMMY, what if people stare at me in the street?" asked eight-year-old Helen O'Toole. "Just wink," advised her mother.

For till the end of April, lively little Helen must lie almost motionless, with a plaster cast around her body and an amazing steel cage over her head, attached to a steel "halo" screwed into her skull. The hardware is designed to stop even the tiniest movement in her neck. She doesn't complain. She knows it will save her life and that she, the first in this country to have had such an operation, is bringing hope to others with the same very rare disease.

Helen's is an inspiring story of courage and dedication: her own, her family's and that of Dr Steven Kopits, a Hungarian orthopaedic surgeon and the only man in the world who has been able to help children like her.

She suffers from Morquio, one of seven muco-polysaccharide diseases (MPS) which are enzyme disorders that have any or all of a range of devastating effects upon bones, physical appearance and physical and mental abilities. There are 350 MPS children alive at any one time in Britain and there is no cure. Their life expectancy is usually at most only 20-odd years, very many are unlikely to grow above three and a half feet tall and they all suffer progressively more handicaps.

Helen lives in North London with her social worker parents, Mary and Charles, and her 10-year-old sister Kate. As Morquio children are unaffected mentally, her disability is in her joints: her wrists are so loose she cannot grip easily, her legs are very knock-kneed and, because she has no "peg" between the top links of her spine, her neck is dangerously unstable. She will never grow taller than she is now, three feet three inches.

Because of her unsupported neck, any fall has meant risk of compression of her spine and total paralysis or death. After two severe falls in 1984, she kept losing sensation in her arms and legs, and her surgeon tried to save her life by taking bone from her hip and wiring it into her neck.

"Helen knew she could have died from another fall if she didn't have that operation. She asked us. We thought it best to be honest because we couldn't hide our fear – she saw it in our faces whenever she tripped," said Mary.

But the operation was only successful in the short term. The bones didn't fuse as hoped.

"When I was told it hadn't worked I thought, either I get very, very depressed or I try to do something," said Mary. So she suggested that the MPS society, a support group for families, should ask the American-based expert surgeon, Dr Kopits, if he would be willing to address a seminar whenever he was next in London.

Her aim was to collect together interested consultants who might be inspired to follow Dr Kopits's techniques to help children here. Christine Lavery, who founded the society after her own son died at the age of seven, was very

enthusiastic. So, to their amazement, was Dr Kopits.

He not only addressed a group of doctors and families of eight Morquio children who were able to come but he offered to do a clinic for each child there and then. "He is such a marvellous, warm man," enthused Mary. "He enchanted them all just by the way he is."

For Mary and Charles, however, the day brought devastating news. "He told us that Helen was very seriously in danger and needed surgery now. We didn't sleep that night, and neither did Christine and Robin Lavery. They went through their accounts and rang Dr Kopits next day to see if we could afford him. But he offered to come back and do the operation for nothing and to teach any interested English consultants at the same time. We were overwhelmed by his generosity and dedication," said Mary.

Dr Kopits's dedication was to be even more astounding. Last December he was leaving his professorial post at Johns Hopkins University Hospital in Baltimore to set up, from his savings, his own clinic for "the little people". Yet he wrote to say he could manage to come over on Christmas Day, arriving on Boxing Day. It was then up to the MPS society to find a hospital willing to loan its theatre.

There began feverish and desperate activity to circumvent NHS red tape. The major children's hospital at Great Ormond Street was unable to make arrangements, so Mary turned to Helen's own consultant at University College Hospital. He was keen, then found that Friday, December 27, was a statutory holiday and only one emergency theatre would be open. It wasn't on.

"It was a nightmare," said Mary. "I took a stiff drink at noon and rang the senior nurse manager in charge of theatres myself. She was so sympathetic once she heard the whole story and made arrangements to bring staff in specially on the Saturday. Once it was fixed, we met with nothing but goodwill."

Meanwhile, families in the MPS society had rallied round to help raise money for air fare and accommodation for Dr Kopits and his nurse. "They were so supportive," said Mary. "Parents who knew their own children were going to die wanted to help one who could live." But, in the event, TWA offered free flights and Trust House Forte provided accommodation at the Waldorf Hotel in London.

Helen spent several hours in theatre. First the steel halo had to be screwed into her skull under local anaesthetic, because her responses were vital for assessing the correct position. Then she was wound into a plaster cast and the steel bars bent to fit over it. Under general anaesthetic next day, three pieces of bone from her legs were grafted on to the top of her spine.

Helen's own consultant and another from the Bristol Children's Hospital were present to watch and now expect to be able to perform the same operation for other Morquio children.

For four months Helen will be unable to sit or stand. "But she is so brave," said her mother. "She just gets on with things and keeps on smiling." The atmosphere at her home is normal and relaxed, much due to the remarkable inner strength and outer ease shown by Mary and Charles and supportive sister Kate. Helen is very much Helen under all her ironmongery and her friends have adjusted to its presence quickly.

The arrival of a special flat wheelchair means she can also now go out into the street – and wink.

There is still a long road ahead. When the halo is removed, Helen will painstakingly relearn how to hold up her head, sit and walk. She will never be able to walk far but her life expectancy – and soon that of many other Morquio children – should now be normal. If anyone has the psychological power to overcome her physical handicaps, she has.

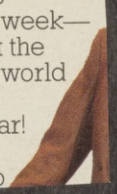
"She knows she will always be a little lady. She has already accepted that."

In gratitude, the MPS society donated £2,500 towards Dr Kopits's new clinic in Baltimore but it always needs money to help families and fund research here. Please write to Christine Lavery, The Society for MPS Diseases, 30 Westwood Drive, Little Chalfont, Bucks.

WOMAN'S OWN

MAY 31, 1986

Features



Helen, you're a real star

Imagine spending five months with your head locked in a metal cage, able to look only up or down, and saying at the end of it: 'I'm getting very good on clouds.' Alan Bestic meets the bravest eight-year-old of all. Photographs by Sally Fear

Tilly the hamster is put on the sofa for her to look at and Helen O'Toole's smile has never been broader. Tilly is her pet, her friend, but he'll never know the measure of her debt to him in the months since Christmas.

By just being there, behaving in a normal, happy hamster fashion, he's captured her spirits whenever they've sunk. He's brought life into days that could have dragged. Tilly has his faults, but being boring is not one of them.

For the last five months, boredom could have been a major enemy for eight-year-old Helen who has a rare genetic condition known as Morquio Syndrome.

She's fortunate in that her life has been saved by Dr. Steven Kopits, a remarkable American surgeon who grafted bone into her neck. But it has meant Helen spending five months encased in a metal frame designed to immobilise her head and neck—and known as the Halo. Since Christmas, she's slept, ate and lived in her Halo and, even more remarkably, accepted the need for it with an understanding beyond her years.

This doesn't surprise her parents, Mary and Charles, at all, because Helen already had an exceptional track record for accepting truths many adults would find difficult to handle.

She already knew, for instance, that Morquio's Syndrome would restrict her life permanently. She would never be able to walk far—a wheel-chair would always be close to hand. And she would always be one of the world's little people—she is only 3 ft. 3 in. tall and because of Morquio she will never grow any more.

The first signs of the condition appeared soon after Helen's birth, but they were so undramatic that Mary and Charles, social workers living in North London, scarcely worried about them. Her muscle tone was not too good. Her limbs were floppy.

But when Helen was 8 months old, specialists found a kink in her spine. Soon afterwards an orthopaedic consultant told her parents he suspected one of the mucopolysaccharide diseases—MPS. The O'Tooles looked them up and were shattered. They cause not only physical but mental deterioration. In some cases, life expectancy is no more than 10 years.

"When tests revealed Morquio's Syndrome we were enormously relieved," said Helen. "We knew it caused no mental handicap and we felt we could cope with the physical problems. It was the prospect of our child losing her mind that we found so frightening.

Knitting: it's easy with Mum's help

Without it, her neck could break in a bad fall. And the long-term prospects were equally grim. Helen's spinal cord would be subjected to chronic wear and tear. Her legs would be affected and eventually she would be unable to walk, then unable to sit and finally unable to breathe. Her life expectancy was estimated at between 20 and 30 years—barring accidents, the simplest of which could kill her.

For three years, however, Helen stayed clear of trouble. Then she fell again and the numbness returned. The family's orthopaedic consultant told them an operation to fuse the vertebrae could protect her but she was too young. Ideally she should wait for three years.

Helen, an astute little girl, inevitably realised there was something seriously wrong. She had listened to her parents constantly telling her not to play on the stairs, not to stand on her head, not to do somersaults. She had seen the fear in their eyes when she fell.

"Naturally she wanted to know why she couldn't do what other children did," said Mary, "so I drew myself to tell her the truth because I felt too sad about it. It was only when I'd managed to accept the situation that I was able to answer her questions.

"But it was hard coming to terms with her future. She stopped growing very soon and I used to feel dreadful when I was thinking of new clothes. She didn't need a lot because she hadn't grown out of what she had already. At eight, she's wearing the clothes of a three or four-year-old. And she soon realised herself that she wasn't growing as fast as other children.

"She used to ask me about it and for a long time I couldn't bring

Working: Helen is still doing well in school

"After that, she asked no more. She came to terms with her future more quickly than I did. She knows now that she's going to be a little lady—a term used in America where they have a 'Little People's Association' for those who have not grown."

There were other problems, too. Helen was knock-kneed. She had floppy wrists and weak ankles. She could not raise her arms above her shoulders. Worse still, when she

'She understood a fall could mean death'

was three years old, she had a fall and couldn't get up—she'd lost sensation in her arms and legs.

Feeling soon came back, but X-rays revealed a frightening flaw in her spine—her odontoid peg, a piece of bone sticking up between the first and second vertebrae of the neck, scarcely existed. It's needed to stabilise the neck.

Learning: a computer is still within easy reach

pictures for her, showing how her neck was vulnerable. I told her she could die if she fell heavily and she understood. I'm sure she would have worried more if I hadn't told her the truth."

But the truth could not protect Helen from falls. Each time she tumbled, the effects were more severe. By the autumn of 1984, the slightest jolt was numbing her arms and legs and a myelogram—an X-ray of her spinal cord—revealed her neck was in an extremely poor condition. The operation was now imperative and, that November, a piece of bone was taken from her hip, put into her neck and wired into position. For a week she knew sickening pain and, in January last year, an X-ray revealed that the bones had not fused.

The O'Tooles, who had taken such a battering through the years, picked themselves up and courageously fought back. For three years they had been members of the MPS Society, a support and self-help group for parents of children with mucopolysaccharidosis. It had been founded by Christine and Robin Lavery whose 7½-year-old son, Simon, had died from Hunter's Syndrome, one of the most severe MPS conditions.

Through the society, Mary and Charles learnt of Dr. Steven Kopits, a Baltimore surgeon who had performed 28 fusion operations on children and adults with MPS. The last 25 had been successful, which meant he was by far the most experienced and successful man in his field. Over the years he had developed a variety of techniques to overcome the problems presented by Morquio patients—among them the Halo—knowledge that could prove invaluable to surgeons in this country.

Mary suggested to Christine Lavery that *(Please turn to page 27)*

the society should invite Dr. Kopits to Britain to address a seminar for surgeons interested in MPS diseases. Christine agreed and so did Dr. Kopits.

"My aim was solely to increase knowledge and understanding of these techniques," said Mary. "But I was amazed when Dr. Kopits offered to hold a clinic for our Morquio children."

He saw eight of them, including Helen. And, in her case, the diagnosis was devastating. Her neck was so severely affected, she needed urgent surgery. Without it her parents were told, she might not live longer than six months.

"Charles and I didn't sleep that night," said Mary. "Neither did Dr. Kopits. Next morning he said he'd

'His wonderful offer gave us new hope'

return to England to operate on Helen and teach other interested surgeons the techniques he had developed.

"It was a wonderful offer because his appointment book was full and that December he was moving into a new international centre for patients with skeletal dysplasias—those with bone malformations, his 'little people.' Yet he was prepared to make time for us if we could organise operating facilities."

That proved to be a frustrating task for Helen's parents. Dr Kopits' only free time was the week after Christmas—he was prepared to leave for London on Christmas Day. But Mary found hospital administrations were not able to be so accommodating.

"That was the worst week of my life. The first one I approached simply wouldn't hear of it. Then our consultant suggested we should try University College Hospital where he operated. I phoned and was told that they'd been forced by financial cuts to admit emergencies only that month.

"Nevertheless, they made us a special case—and then I learnt that the day we'd set aside for the operation, Friday, December 27, was a staff holiday. It would have to take place the following week. And that was too late for Dr. Kopits.

"By that stage I couldn't stop crying. It was one of the worst days of my life. Helen was deteriorating, beginning to crawl rather than walk. If she stopped walking, she'd never walk again then she would stop sitting and breathing and then she would die.

"Our consultant has given us magnificent support over the years and suggested that I phone the Nurse Manager in charge of operating theatres at University College Hospital. I was shaking so much I had a whisky to steady my nerves. Then I phoned and she said she couldn't manage the Friday but could organise staff and theatre for the following day."

On December 27, the Halo was screwed into Helen's skull under local anaesthetic. Early next day the operation began and lasted over seven hours. Somehow this

'Two blinks for yes and one for no'

exceptional little girl managed a hello for her parents as she was wheeled from the theatre to the intensive care unit.

"It was her last word for a while," said Mary. "She was put on a ventilator and from then on she 'talked' to us by blinking . . . two blinks for yes, one for no. I sat with her for the next two nights."

Helen weathered the operation with her rare brand of courage and then faced the long months ahead, caged in her Halo. The frame could be moved so that she could lie on her tummy and look down or on her back and look up. "I can't see sideways," she said to me one day, "but I'm getting very good on clouds."

A teacher called to give her tuition and, by April, she was ahead of many of her classmates. A physiotherapist treated her regularly. Once a week her friends Catherine and Lucy came round to play with her and Tilly the hamster was never very far away. And, naturally, she longed for the day when the Halo would be unscrewed from her skull. But she knew that there would be no immediate return to her normal way of life. She would have to learn to stand and walk again. No longer, however, would there be the fear of death from a stumble, a normal lifespan lay ahead of her.

Mary and Charles, also facing this new era, look back in gratitude. "Our friends are marvellous, particularly those in the MPS Society," said Mary. "There were times when I needed to be with families in a similar position to ours . . . often worse than ours. With them I can refuel. With those who have able-bodied children we have to explain so much. With MPS members we can speak in shorthand.

"MPS has helped Helen, too. She used to ask if she was the only one in the world with Morquio. Since we joined the society she has learned there are many others like her and she has made friends with about 10 of them, so she's not an island any more. And neither are Charles and I.

"And there's another reason why MPS has become an important part of our lives. I know now that I can do something to help promote knowledge of these rare conditions. Helen knows she can help, too—and that helps her." ●

For further information on MPS, contact: Mrs. Christine Lavery, The MPS Society, 30 Westwood Drive, Little Chalfont, Bucks.

Playing: a game of cards with Mum

Eating: trying not to spill the beans

San Mateo THE TIMES Wednesday, July 23, 1986

THE DAVID NORRISES
(Laurie Wolfe)

American Medical
NEWS

AUGUST 15, 1986

Defeating dwarfism

Orthopedic surgeon Steven E. Kopits, MD, has devoted his professional and much of his personal life to dwarfs. See page 3 for a story on the physician who specializes in the disabling disorder of dwarfism.

International Center for Skeletal Dysplasia

STEVEN E. KOPITS, M.D.
Director

Dear Donna,

Greetings from the East. It was such a delight to meet on your recent visit. I promised to send you some information about England, and here it is.

Mary O'Toole is the Mother of Helen who is the young girl in the enclosed article. She had recently called here and is so happy you are coming to England. Their address is:

8 Elmhurst Avenue
East Finchley
London N2 OLT England

Their telephone # is London, England 01-444-8461

I know you will have a wonderful time in Europe and in England.

Christine Lavery founded the Society
in England and will I'm sure
be most anxious to meet you also.

Have a wonderful time.

Sincerely

Elizabeth Albert

Saturday, Dec. 6, 1986 THE TIMES San Mateo—B1

Peninsula

Town rallying to help sick girl

By BILL HURSCHMANN
Times Staff Writer

BRISBANE —An 11-year-old girl suffering from a rare bone disease is in desperate need of corrective surgery, and the community is rallying behind her. Nicky Bell has diastrophic displasia, a genetic bone disease that has made her a dwarf, with all the bones in her tiny body twisted.

A series of operations that could free Nicky from her wheelchair and allow her to use a walker or cane has been performed successfully several times by Dr. Steven Kopits at Johns Hopkins University in Baltimore, Md., and that's where Nicky's mother, Marlene Bell, would like to take her next year.

The catch is money.

Nicky, a fifth-grader at Lipman Intermediate School, is covered under a couple of insurance plans, Kaiser Permanente and Shriner's, but neither can perform the necessary surgery nor pick up any of the costs if it is performed outside of their facilities.

But that hasn't stopped Mrs. Bell from fighting on all fronts for two years now to garner support and raise funds for her little daughter.

The Brisbane Eagles held a Halloween night fund-raiser which netted $4,000; Nicky's classmates decided to contribute several hundred dollars from fund-raisers into the coffers for her operation.

But that leaves the fund, established at the Bank of America branch in Brisbane, well short of total amount needed.

The initial operation would be on Nicky's hips, which today are pointing out and twisted, making it impossible to link with her ball joints. Then, two weeks later, similar surgery would be performed on her knees, followed by her ankles and, finally, her feet.

She will require up to four months of hospitalization.

The surgery won't relieve the arthritic pain Nicky suffers, and, in fact, doctors have told Mrs. Bell it could get worse. "But it could get worse no matter what, too," she says.

This year is Nicky's second at Lipman School; she first tried to attend Brisbane Elementary, but the difficulty she had in getting up steps caused her hips to swell severely and she transferred to Panorama School in Daly City which has no stairs. Lipman in the interim had a couple of ramps built so Nicky could maneuver around campus.

"Nicky doesn't really have a handicap," says Lipman Principal Marilyn Marino, referring to Nicky's refusal to let her wheelchair inhibit her.

for our children." That fight, she says, has resulted in calls to the White House, in pleas for help to Easter Seals and March of Dimes, to Rep. Tom Lantos and former Assemblyman Lou Papan to hospitals up and down the West Coast.

"I've run up against stone doors for years," she says.

Mrs. Bell admits the fight has been difficult. "When you're fighting something like this, you feel so alone. It's very frustrating and you get real depressed. But my friends say just wait for the benefit, and it all with be worth it."

"Her personality is such, kind and loving, that she adds a lot to our student body."

Other children, Marino says, become more aware of handicaps because of Nicky, and subsequently become more thoughtful and caring.

"All the students liked the idea (of donating money to Nicky)," adds student body President Michael Frias. "We all felt like it was a good idea to help her out."

"There's got to be help out there somewhere," says Mrs. Bell, "after all, we're fighting

Times Photo by Scott Buschman

Nicky Bell talks with classmates at Lipman School in Brisbane.

TOP OF THE WEEK

Hormones

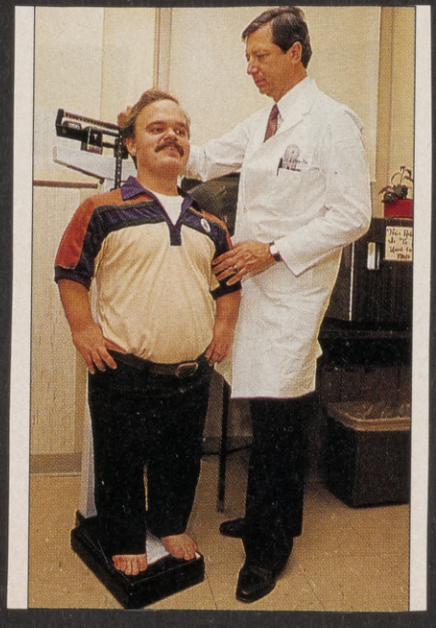
The pituitary, the master gland, helps make basketball stars, while the adrenals help the body gear up for fight or flight. Pheromones—which are not hormones but work like them—may help the sexes attract each other. These mysterious chemical messengers maintain the exquisite balance of being. **Society: Page 50**

Newsweek

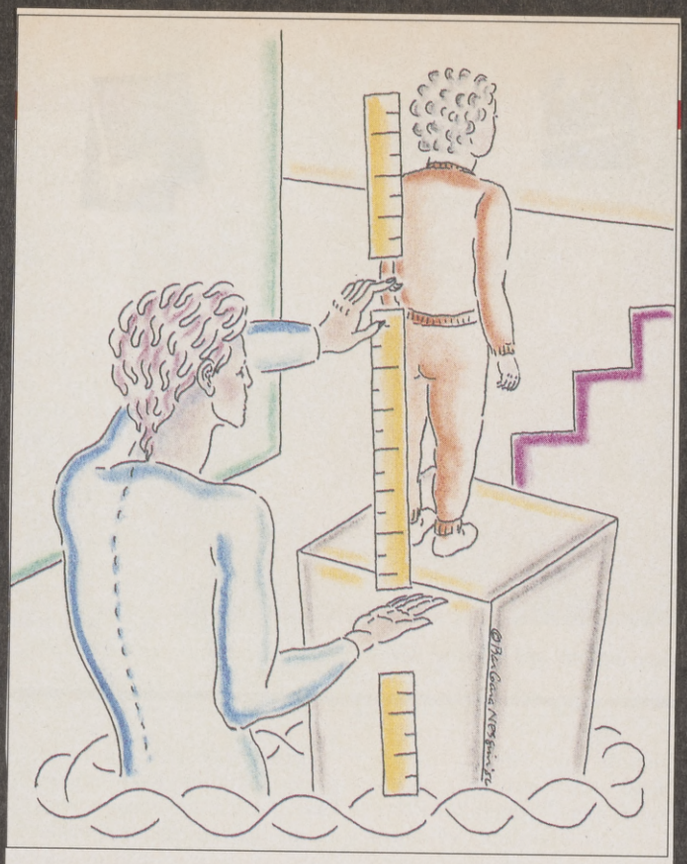
January 12, 1987 : $2.00

HORMONES

How They Affect Behavior, Growth, Sex and Health



PITUITARY
produces growth hormone; insufficiencies can cause dwarfism



GROWTH hormone poses a potential ethical question: should we try to raise only tall children?

Giants, Dwarfs and Cancerous Cells

A pro basketball team is a sports fan's delight, but to endocrinologists it also provides dramatic evidence of hormonal differences. Human-growth hormone, GH, is produced by the pituitary and is the master regulator of growth throughout childhood and puberty. Too much could make for seven-foot centers; too little could create dwarfs. Here, too, man has not had to settle for what nature provides. Until recently, GH supplements had to be extracted from the pituitaries of human cadavers and were in short supply. But since 1985 genetic engineering has allowed GH to be made in vast quantities, and therein lies a potential dilemma. Should short children with low GH concentrations be administered growth hormone? And what about those just a bit below average whose parents associate stature with success?

Prescribing growth hormone can be risky. While stimulating elongation of the bones, it also speeds their maturation. As a result, children given supplements "may just run the developmental program faster," warns Dr. Gordon Cutler of the National Institute of Child Health and Human Development, and may end up just as short as they would normally. Too much GH can also theoretically cause some forms of diabetes and in rare cases make the thigh bone slip out of its pelvic socket, which occurs when bones grow too fast. Overactivity of the pituitary can cause a sudden increase in production of growth hormone in adults. The result is acromegaly, a disease in which the face and forehead suddenly start growing again, with grotesque results.

To date, Genentech and other bioengineering firms that make synthetic GH have limited the distribution to doctors who specialize in treating dwarfism and other specific disorders. "You can't just go out and get it at your corner drugstore," says Dr. Boris Senior, chief of pediatric endocrinology and metabolism at Tufts-New England Medical Center in Boston. Still, Genentech's growth hormone, marketed under the name Protropin, has found a $45 million annual market for treating an estimated 15,000 children who suffer from hypopituitary dwarfism, and it may eventually have other applications as well—healing wounds, fractures and certain states of malnutrition and helping children who don't grow as a result of kidney problems or chronic heart disease.

While GH stimulates the growth of the entire body, hormonelike growth factors stimulate the growth of particular kinds of cells. As such, they may play a role in human cancers. Stiles and his associates at the Dana-Farber Cancer Institute are now looking for substances that might *inhibit* growth factors by altering the genes involved in their synthesis and action. In particular, they are studying PDGF, platelet-derived growth factor, which stimulates certain cells to divide and form scar tissue and may encourage other cells to divide inappropriately, contributing to the growth of a cancer. "What we're asking is whether, in nature, there is not only an accelerator but a brake pedal as well," he explains. "In endocrinology, for every yin there is a yang."

Hormone research has already paid off dramatically in the treatment of breast cancer, a disease that takes the lives of some 38,000 American women each year. Years ago it was learned that estrogen can fuel the growth of tumors in some women, and removal of the pituitary, ovaries or adrenal glands (which also produce estrogen) became a standard therapy. But gland removal is drastic treatment. Without the adrenals, for example, patients must take cortisone for the rest of their lives, since without the cortical hormones produced by the adrenals, patients could not endure any kind of stress and would die.

Scientists are exploring other forms of hormone therapy, including drugs that block the manufacture of estrogen in the adrenals. These antiestrogen drugs para-

THE TIMES Wednesday, Feb. 4, 1987

A Soviet procedure has been brought to the U.S.

Pulse of the Peninsula

By Janet Parker Beck, Times Staff Writer

BURLINGAME — In just two months, Jaime Barrett's left leg has grown more than two inches, nearly catching up with the length of her right leg.

The 10-year-old Burlingame girl is the beneficiary of a Stanford University physician's determination to bring a bone-lengthening procedure developed in the Soviet Union to the United States. Jaime, according to Dr. Eugene Bleck, is apparently the first patient in the United States to undergo the experimental operation.

In Jaime's case, the procedure performed at Stanford on Nov. 25 corrected the long-term effects of a birth defect that resulted in her left leg being 2.8 inches shorter than her right leg.

Jaime, who tolerated a significant amount of pain and frustration to accomplish the goal of nearly even leg lengths over the past two months, is confident that it's all been worth the trouble.

"Sometimes I wonder if I'll regret all this," concedes the Our Lady of Angels School student who will have a home tutor for the remainder of the school year. "But it's getting a lot easier and now I know I won't have to wear a big lift on my left shoe anymore."

MaryAnne and John Barrett thought about their daughter's options, including a more traditional leg-lengthening procedure, before they agreed to the experimental operation. Bleck has been treating Jaime since she was born and, "His word was good enough for us," MaryAnne Barrett declares.

Jaime's birth defect, called congenital pseudoarthrosis, caused a forward and outward bowing of her left tibia, the large bone between the ankle and the knee.

When Jaime was 5 months old, Bleck, chief of the Division of Orthopedic Surgery at Stanford School of Medicine, operated to move a piece of her fibula, the slender outer bone, behind the tibia as a "strut" to strengthen the larger bone. Bone fractures are common in children with these defects and the operation was needed to prevent breaks from occurring, according to Bleck.

Jaime, in fact, never suffered a broken leg during her 10 years of growth.

For several years, Jaime wore a leg brace and, later, a shoe lift. But the youngster still played Bobby Sox softball for one season and even tried gymnastics ("I got bored") for three months.

Bleck had warned the Barretts several years ago that Jaime would need another operation by the time she was 9- or 10-years-old to prepare for her pre-teen growth spurt. According to Bleck, the mean age for girls to stop growing is 13.75 years so it was essential that the major lengthening operation be performed now.

The traditional Wagner method (named after its developer) of lengthening a leg involves cutting the bone through the inner marrow in the middle of the shaft and placing two large screws or pins above and below the cut.

A month later, a bone graft and metal plate insertion is necessary to completely fill in the gap and strengthen the leg.

But Bleck says the Wagner method would have been somewhat "risky" in Jaime's case and probably would not have increased the length of her leg by more than about 1½ inches.

On a trip to Europe in 1985, he observed the results of a procedure developed by Soviet physician Dr. G.A. Ilizarov. The Ilizarov operation is becoming popular among French and Italian orthopedic surgeons, Bleck says.

According to Bleck, 222 cases in which the Ilizarov procedure was used have been reported by 50 French surgeons. Bleck was impressed by their results.

The newer procedure involves, he explains, making tiny cuts through the cortex — the hard outer part — of the bone near its upper end, then running fine wires through the leg and bone. The wires are attached to metal rings encircling the leg.

During the recovery, the patient turns the nuts on the metal rings four times a day to slowly pull the bone apart one fourth of a millimeter — around one-hundreth of an inch — at a time.

In cutting only the outer layer of bone, leaving the nourishing marrow and blood supply intact inside, new bone formation fills in the gaps in the cortex over time, Bleck says.

A French surgeon, Dr. Sylvain Terver, demonstrated the Ilizarov procedure at Stanford during a visit in November. He had been a visiting scholar at Stanford and Children's Hospital at Stanford in 1979 and 1980.

Although Soviet surgeons have

Times Photo by Ray Zirkel

RECOVERING AT HOME

Jaime Barrett is looking forward to the day she won't need to depend upon a shoe lift to walk normally.

been developing the technique since 1952, it needed to be perfected before it could be offered elsewhere, Bleck says. In addition, the Soviet's didn't publish the results of their operations in other than Russian publications which weren't readily available, he says.

"The Europeans are always apt to try new things quicker than we are because we have all sorts of constraints," Bleck says. "Many people feel it's not worth it to fight those constraints."

Bleck, however, felt otherwise.

Bleck purchased the equipment for the Ilizarov operation using funds donated to his department. Working through the Stanford Human Subjects Committee, permission was obtained from the U.S. Food and Drug Administration to perform the operation.

Bleck assumes it was the first Ilizarov leg-lengthening operation in this country because the FDA had no record of anyone else asking for approval. The equipment, Bleck says, is available only from a French supplier.

After the five-hour surgery, Jaime spent 12 days in the hospital. She recalls screaming when anyone even approached her mending leg during those early days.

"It was scary," MaryAnne Barrett says with a grin. "She yelled so loud, we thought, 'This can't be OUR daughter!' "

"I didn't even want to look at it," Jaime recalls. It was nearly two weeks before she felt comfortable about peeking at the metal device wrapped around her leg and attached with wires through the bone.

At home, Jaime and her mom carried out the pin-turning procedure — which is now completed — four times every day. Her follow-up care also includes spending 16 hours a day with her leg in a perpetual motion machine designed to keep her knee joint lubricated. Jaime also is required to exercise her leg three or four times a day for 20 minutes.

The physical therapy is aimed at strengthening and "fattening up" her left leg, stimulating the production of calcium to promote bone growth, and achieving the ability to fully straighten her leg.

Jaime props the metal hoops on pillows at night in her bed and she's learned — somewhat reluctantly — to sleep on her back. "I use to move all over the place in my sleep," she declares.

A few weeks ago, the young patient was able to begin putting weight on her left leg. She's now getting about with the help of crutches.

Jaime spent another five days in the hospital in mid-December for a second, shorter procedure to stretch her Achilles tendon — the heel cord. The cord had atrophied because Jaime has walked with her left leg on tip-toe for most of her young life.

Her leg is now 2.3 inches longer than it was in November. Bleck says that over the next two months, she will continue to wear the apparatus as the bone continues to fill in the gaps. This will enable her leg eventually to withstand a full weight-bearing load, he says.

The greatest dangers of a leg-lengthening operation, the surgeon explains, are infections from the pins, overstretching of nerves and arteries, and stiffness of joints from compression of cartilage. None of those potential complications, however, has been of significance in Jaime's case. With the Wagner method, infection is more likely because the pins are so much larger than the Ilizarov wires, Bleck says.

The Wagner method, however, is still useful for certain types of bone-lengthening, he adds.

According to Bleck, the Ilizarov method has other possible applications. Surgeons might use it to repair open fractures of the tibia or replace bone loss from injury or tumor removal, he says.

A few European surgeons, he notes, have been using the procedure in a controversial effort to lengthen the legs of children afflicted with dwarfism. Bleck says such a procedure is complex, risky, difficult on the patient and expensive. Numerous operations would be required. Stanford will not be offering the Ilizarov procedure for treatment of dwarfism.

The Barretts didn't realize their daughter was going to make medical history in this country until it was nearly time for surgery. Watching what Jaime has been through has "made us all stronger," MaryAnne Barrett says.

"We've learned a lot about the fibula and the tibia over the past 10 years," she says.

Jaime is looking forward to returning to school in the fall and, hopefully not too long after that, playing softball again.

The young patient has ventured out on a few shopping trips since the operation but she's felt a bit uncomfortable when people have stared at the metal device secured to her leg. It's taught the youngster a valuable lesson.

"I'm never going to stare at anybody else after this," she says with a smile.

Peninsula

Saturday, May 16, 1987

Lions Club raises money for local girl's operation

By BILL HURSCHMANN
Times Staff Writer

BRISBANE — The Brisbane Lions Club will hold a fund-raising dinner Sunday for Nicky Bell, a 12-year-old Brisbane girl who needs money for an operation to correct a rare bone disease that has left her a dwarf, twisting the bones within her body.

Nicky, a Brisbane native, is a fifth-grader at Lipman School, a member of the school band and maintains a straight-A average. Her mother, Marlene, daughter of a former Brisbane mayor, has been trying for months and months to raise the necessary $150,000 needed for an operation that will help Nicky walk.

Nicky suffers from diastrophic displasia, a rare and crippling disorder that has deformed all of her bones and has confined her to a wheelchair since she was 4.

The Lions, said club member Lee Panza, are holding the benefit dinner because "we believe in Brisbane as a community where people still care for one another, and we're asking for your support for an exceptional little girl."

The dinner will be held at the Community Center, 250 Visitacion Ave. A donation of $20 per person is suggested, and two entrees are available, ham steak Hawaiian with champagne sauce or roast prime rib. There are two dinner seatings, 4 p.m. and 6:30 p.m.

State Sen. Quentin Kopp, I-San Francisco, and San Mateo County Supervisor Mary Griffin from Millbrae are expected to attend, Panza said.

Although Nicky could once walk, she never will again unless she has the surgery, Panza added. Nicky must undergo a series of at least eight major operations to reconstruct her legs and hips, and it must be done within the next few months before her bones harden and set in their permanent shape.

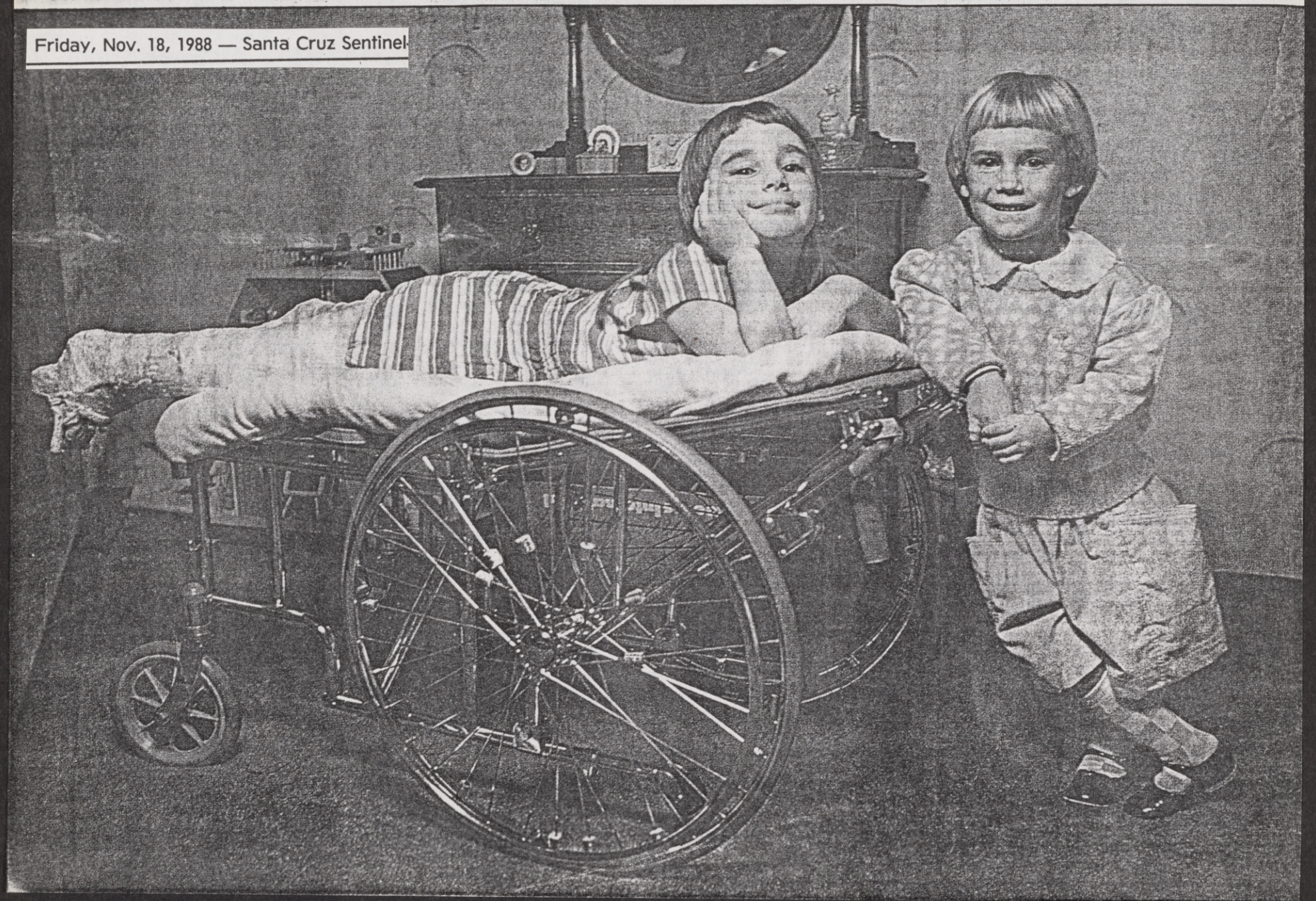
Dr. Steven Kopits, who heads the Skeletal Displasia Center at Johns Hopkins University in Baltimore, the only one of its kind in the world, will perform the surgery for $150,000, and Nicky's medical insurance will not cover it.

"For a working single parent this (amount of money) is simply out of reach, particularly in the short time remaining," Panza said. "So Marlene has turned to us, her Brisbane neighbors, for help. The outpouring of concern and generosity has been heartwarming, and has given both Nicky and Marlene hope that they will succeed. But they're still far short of the sum they have to raise."

Anyone wanting to donate to Nicky but unable to attend the fund-raising dinner can send donations to the Nicky Bell Trust Fund Account at the Brisbane branch of the Bank of America, 70 Old County Road.

ittle girls face big challenge

Friday, Nov. 18, 1988 — Santa Cruz Sentinel



Baltimore doctor offers hope to sisters with rare type of dwarfism

By TOM LONG
Sentinel staff writer

EDITOR'S NOTE: The parents of Jennifer and Danielle asked that their last names not be used in this story.

JENNIFER said her legs hurt. So her parents, Michael and Karen, did what any good parents would do. They took their 3-year-old daughter to a pediatrician.

"The first doctor dismissed it as growing pains because he couldn't find anything wrong," Karen remembers. "He told us to bring Jennifer back in three months if it was still bothering her."

So they did, and the pediatrician referred the family to an orthopedist. The orthopedist took X-rays of Jennifer's legs and found something wrong — but he didn't know what. His suspicion was aroused. He referred Jennifer to a geneticist.

The geneticist told the family that Jennifer had pseudo achondroplasia — a rare type of dwarfism.

Two days before the geneticist delivered his diagnosis, Karen found out she was pregnant with the couple's second child.

The geneticist reassured Michael and Karen.

"They said, 'You don't have anything to worry about,' " Karen says.

According to the geneticist, only about one in 100,000 children are born with pseudo achondroplasia, the only form of dwarfism that isn't apparent at birth.

"The chances against it happening twice would be astronomical," Karen remembers the doctor saying.

Both parents had come from average-size to tall families. Neither had any history of dwarfism in their families.

Six years ago, Michael and Karen had their second daughter, Danielle.

Like her older sister, she was adorable.

And, like Jennifer, Danielle had pseudo achondroplasia.

DISCOVERING that Jennifer was a dwarf did nothing to make the pain in her legs go away.

The pain was caused by abnormal growth patterns in the bones in Jennifer's lower half. Pseudo dwarves have virtually normal development of the upper torso and head, but their legs and hips undergo the same confused growth patterns that have crippled many dwarves.

Sometimes the legs grow bowed, other times the knees grow in toward one another. In Jennifer's case, her legs were developing a "wind-swept" appearance, both radically drifting to one side.

So Michael and Karen and Jennifer continued going to doctors.

They went to university medical centers. They went to famous surgeons up and down the West Coast. They talked to a variety of orthopedic surgeons and specialists. And they noticed something: There wasn't much agreement on what to do.

"We were getting conflicting information on how to treat her," Karen says.

"Some of the doctors wanted to use braces," Michael remembers. "Some wanted to operate on one leg and wait and see what happened and then operate on the other leg."

"We just knew we didn't want Jennifer to end up being a guinea pig," Karen says.

The search for treatment lasted about two years. Those were lonely years.

"We had no support group. We didn't know where to go, what to do," Karen recalls. And then the family discovered Little People of America.

LPA is an organization run by and for dwarves or, as they prefer to be called, little people. It has thousands of members nationwide and a healthy membership in the San Francisco Bay Area. Michael and Karen met adult little people and normal-size people who also had dwarf children. Jennifer and Danielle met other kids their size.

Most important, the family heard about a doctor. A doctor who helps little people.

DR. STEVEN Kopits is probably the only surgeon in the world who specializes in treating little people. He has worked his miracles on more than 700 of them since 1970. He lives in Maryland and is based at the Johns Hopkins University of Medicine in Baltimore, but travels often to LPA conventions to meet prospective patients.

When Michael and Karen first heard of Kopits, they wrote him a letter describing Jennifer's condition and asked if he could help them.

Kopits wrote them back one word — yes.

The family journeyed to an LPA convention in Seattle to meet Kopits. Danielle, who had not yet been diagnosed, walked into the room first. Kopits looked at the little girl, whose parents were sure she was not a dwarf, and thought she was the patient.

That's how the family learned that Danielle also had pseudo achondroplasia.

Girls/ Surgeries won't stop dwarfism

Continued from Page D1

And while the parents were stunned by that revelation, they immediately knew that Kopits was not like all the other doctors they had seen. He was not guessing. He was not stymied.

He knew.

"He's the only doctor we talked to who understood what we were talking about and we understood what he was talking about," Michael says. "The other doctors didn't seem to have the knowledge or the ... camraderie."

Since that time, both girls have been operated on by Koptis at Johns Hopkins. Koptis performs surgery on dwarf children at the end of growth spurts, removing sections of their bones that have grown at a quicker rate than others. He tries to line up the growth rates for the next spurt. By the time his patients are full grown — which for most dwarves means a height of under 4½ feet — the surgeries will hopefully have balanced their lower body growth.

"His goal is to have them able to go to college in another state," Michael says. "Because that means they're able to be totally on their own."

RIGHT NOW, Jennifer is in a cast from the waist down. She wheels herself about on a wheelchair-table contraption. She had her last surgery Oct. 1 and will remain in the cast until early December, when she will fly back to Baltimore so that Kopits can unveil the new configuration of her legs.

Lying flat on her stomach, she still manages to sing in the choir for a Santa Cruz church while waiting to get back on her feet.

Danielle is in kindergarten and will happily point to the scars on her legs left from Kopits' first surgery on her. She bounces about like any 6-year-old, beaming a pixie smile, full of enthusiasm for life.

There are many other aspects to this family's story. Michael and Karen's economic struggle to get their kids the best treatment possible, the wrestling matches with insurance companies, the stares when the family goes out for dinner at a restaurant. There's the emotional balancing act involved when normal kids live in abnormal bodies.

But the facts are these — Danielle is 38 inches tall now, Jennifer is 44.

Kopits estimates that when Jennifer is full grown she will be about 4 feet, 4 inches tall. Danielle may be a few inches taller.

They are little people. With big hearts. And if all goes well, they will be able to walk straight and tall for the rest of their lives because their parents refused to give up the search for the right doctor.

Any one wanting to contact Little People of America may do so by writing to at P.O. Box 633, San Bruno, Calif. 94066.

Benjamin S. Carson, M.D.

"PLEASE OPERATE ANYWAY."

A YOUNG DOCTOR FACES HIS TOUGHEST CHALLENGE

The mother and father standing before me in the consulting room would not believe there was no hope for their four-year-old son. They had brought him to Johns Hopkins from their home in Georgia, where he had been diagnosed as having a malignant tumor of the brain stem, that knoblike cluster on top of the spine through which all brain impulses flow. The little blond boy was paralyzed, comatose; his blue eyes gyrated sightlessly.

I suffered with the parents. I had three small sons of my own. Yet I had studied the X-rays revealing the dark, ugly mass and discussed them with the radiologists and others on the staff. "I'm so very sorry, Mr. and Mrs. Pylant," I said, "but there is no way in which we can encourage you."

The mother's chin quivered. "That's what they told us in Georgia, doctor, but the Lord led us to bring Christopher to Baltimore because He made it plain there was a doctor here who could help him. We believe you are that doctor."

"It's not in the mind of the Lord to let our little boy die, doctor," broke in the father, nervously twisting the brim of his fedora. "Will you please operate anyway?"

In the face of such faith what could one say?

In the face of such faith, what else could I say?

The next morning after my regular Scripture reading, I prayed, asking God to guide my hands and mind in the complicated operation facing me. In communing with Him, I thought about all the unbelievable things He had wrought in my life since I was growing up in Detroit's inner city.

My mind drifted back to those days when my mother raised my older brother, Curtis, and me all by herself.

She prayed every day for Curtis and me and on Saturdays took us with her to the Seventh-day Adventist church. I loved the stories about the prophets and Jesus and His healings. And when I heard how the mission doctors helped people in far-off lands, I vowed right then and there to become a physician.

I told my mother about my dream as we walked home along the glass-strewn sidewalks one night. Aware of the hopeless-looking men standing in doorways and the squad car racing up the street, she stopped and put her hands on my shoulder. "Benny, if you ask the Lord for something, *believing* He will do it, then He *will* do it."

But my mother was also well aware of my poor marks in elementary school, and she proceeded to let me know it would also take a lot of work on my part.

"You will never become a doctor if all you do is watch television," she said one morning as she snapped off "The Three Stooges." "You and your brother had better start reading something."

She insisted that we read at least two books a week. "Don't you touch that knob, Benny," she'd ordered if ever she caught me reaching for the TV. "Read your book."

And so I did, and the more I read, the more interesting books became. Before long I was devouring them.

Within two years I rose from the bottom of my class to the top. My good marks won honors in high school, which earned me a scholarship to Yale University, then the University of Michigan Medical School, and eventually helped me realize another long-held dream, a staff appointment at Johns Hopkins Hospital.

But how easily none of these things might have happened.

One of the problems I had as a kid was my violent temper. It was so severe that when it exploded I'd even attack others with a rock, brick, or anything else at hand. No matter how hard I tried to control it, my temper would snap like an old rattrap.

PHOTO: DENNIS CREWS

One day when I was 14, a boy in our neighborhood was tormenting me. Suddenly everything flashed red. Snatching a big camping knife, I lunged fiercely at his stomach.

Crack! The steel blade snapped as it struck his heavy metal belt buckle.

As the boy fled in terror, something also snapped in me. I was shocked at what I had nearly done: I might have *killed* that boy. Sickened, I stumbled home, where I shut the bathroom door and slumped on the porcelain tub, staring at the wall.

How could I help myself? I knew that my temper was out of control. Something had to be done.

I felt so sorry for the Pylants, who believed so strongly in something so hopeless.

At that time in church we'd been reading the book of Proverbs. It was my favorite book—as it is now. As I sat on the edge of the tub, some of Solomon's words began to form slowly in my mind: *He that hath no rule over his own spirit . . .*

I couldn't help but feel that there was meaning for me in those words: *He that hath no rule over his own spirit is like a city that is broken down, and without walls* (25:28).

They *were* meant for me; I knew now for certain that I had to get control of my spirit. If I didn't I'd end up in jail, or dead.

"Ask the Lord, Benny, *believing* that He will . . ." Mother had said again and again.

Right then and there I knelt on the bathroom floor. "Oh, Lord," I prayed, "take away my temper. I know, I *believe* You will."

And He did. There wasn't anything gradual about it at all. The Lord took away my temper, just like that. Whenever I'd feel it begin to boil, it would somehow simmer down as if someone had turned off the burner. I was in awe at what had happened to me, and I remained so.

When the time came to operate on little Christopher Pylant, I looked at my hand, the one that had wielded the knife that fateful day, and I gave thanks that it was about to guide what I hoped would be a lifesaving scalpel.

That morning in the operating room, after I'd opened the back of Christopher's little shaven head, it was just as I expected: malignant dark tumor everywhere. I couldn't even see the brain stem, which evidently had been consumed by the cancer. Without the brain stem, there is no real life.

We excised as much of the tumor as safely possible, closed the incision, and had the boy taken to the intensive-care unit.

As I stepped into the waiting area, the parents rose with expectant faces. I didn't have the heart to tell them there was no hope, but I had to.

"I'm sorry we couldn't help your son," I said. "I know you both have prayed, and I have prayed too. But sometimes the Lord works in ways we don't fully understand."

The mother and father did not flinch. Both still maintained that earnest look of conviction.

"Doctor," the father said as his wife nodded in agreement, "the Lord is going to heal our son. We're standing on His promise."

Taking a deep breath, I could only add, "Your faith is admirable."

I felt so very sorry for the Pylants, who believed so strongly in something so hopeless. What I had seen in Christopher's brain was irrefutable.

However, three days after the operation something strange happened. Though Christopher was still comatose, his eyes began focusing, and his physical movements began improving.

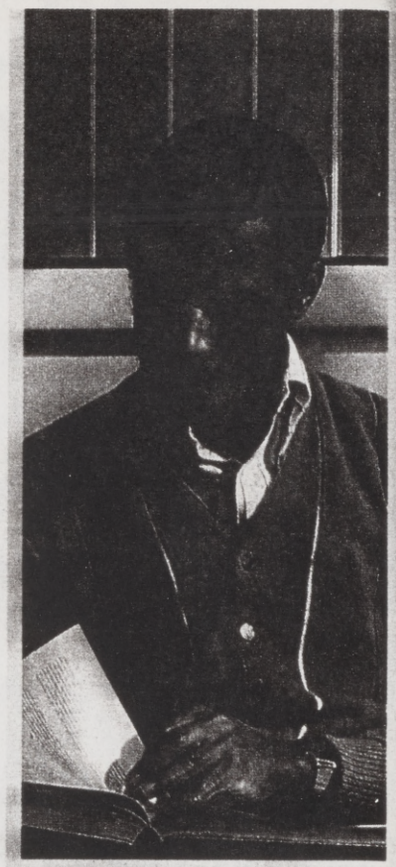
"Let's repeat his CT scan," I said, a peculiar feeling going through me.

As I studied the scan printout, I was amazed to see a tiny threadlike remnant of brain stem.

We had to go back in.

The next day, Christopher was back in surgery, and I was again working on that discolored malignant tissue with scalpel and forceps. But where it had seemed so impossible to define planes before, I was suddenly able to lift the mass away in layers. I became excited. A nurse wiped perspiration from my brow as I worked. At last, after cleaning out all the crevices, there it lay, the healthy gray brain stem, intact, but flattened and distorted.

Within a month our patient was ready to leave the hospital. With Christopher smiling up at us, his parents and I thanked the Lord together. As they walked out of the hospital, the glory shone on their faces, and I heard my mother telling me once again: *"If you ask the Lord for something* believing *He will do it, then He will do it."*



The Rest of the Story

Today, four years later, Christopher Pylant is active and healthy.

Dr. Carson was the primary surgeon in the dramatic and successful 22-hour operation in September 1987 separating the West German Siamese twins joined at the head. He has also performed a number of hemispherectomies, a rare and critical operation in which half of a child's brain is removed to keep him or her from deteriorating or dying of a rare seizure disorder, Rasmussen's encephalitis, which initially affects only one hemisphere of the brain. Those children have recovered almost all faculties, since the remaining half of the brain evidently takes on the functions of the other.

Dr. Carson (whose middle name is Solomon) reads Proverbs "for wisdom" in the morning and at night.

He graduated magna cum laude from high school, attended Yale University and the University of Michigan School of Medicine, interned and took residency at Johns Hopkins, and was senior neurological registrar for one year at Sir Charles Gairdner Hospital, western Australia's major neurosurgery center. He returned to Johns Hopkins in 1984 and at age 33 was made its director of pediatric neurosurgery.

Bell Travel System Spring 1989

Super new Mobility 2000 wheelchair allows disabled travelers to sightsee as never before.

This Wheelchair Will Take You Traveling!

Do you know someone who's confined to a wheelchair yet who loves to — or longs to — travel? A British company named Mobility 2000 has invented what it calls a climbing wheelchair which promises to make sightseeing almost normal for disabled people.

According to the company, Wheelchair 2000 is a versatile, compact, battery-operated indoor/outdoor chair that has a small turning radius, can climb inclines, curbs, steps and full flights of stairs, and can navigate rough or soft ground. In addition, the chair can be folded into a compact cube that will propel itself into a van or station wagon; the seat can be raised and lowered; it reclines at the touch of a button; and it can travel at up to four miles an hour.

For a list of the company's U.S. sales agencies, contact Mrs. S. R. Hester, Mobility 2000 (Telford) Ltd., Telford Industrial Centre, Stafford Park 4, Telford, Shropshire TF3 3BA, Great Britain. Telephone: (0952) 610329.

Growing up short is no small matter

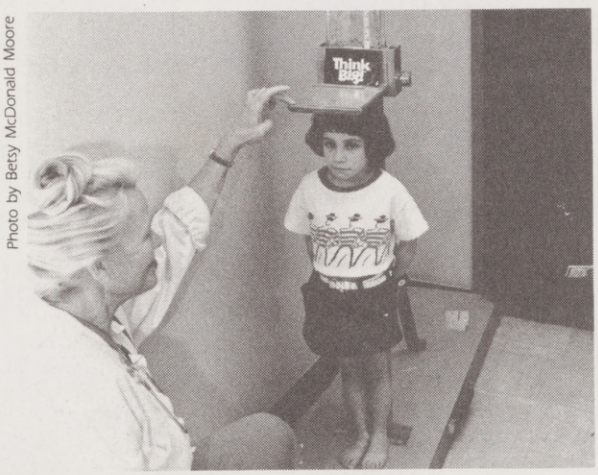
By Betsy McDonald Moore

Eight-year-old Sam can barely reach the drinking fountain on his tippy toes. Unlike his fellow third graders, he has to use the same footstool that the kindergartners use to get a drink. He can hear his classmates giggling. The color slowly creeps up his neck to his face until his cheeks burn from the heat of embarrasment.

Sam suffers from growth hormone deficiency which makes him noticeably shorter than other children his age. It is

Continued on 2

Photo by Betsy McDonald Moore



Think big: Pediatric endocrinology nurse Lois Roundtree, RN, uses a stadiometer to measure 7 1/2-year-old Sarah Garcia. Sarah hopes to reach three feet by this summer.

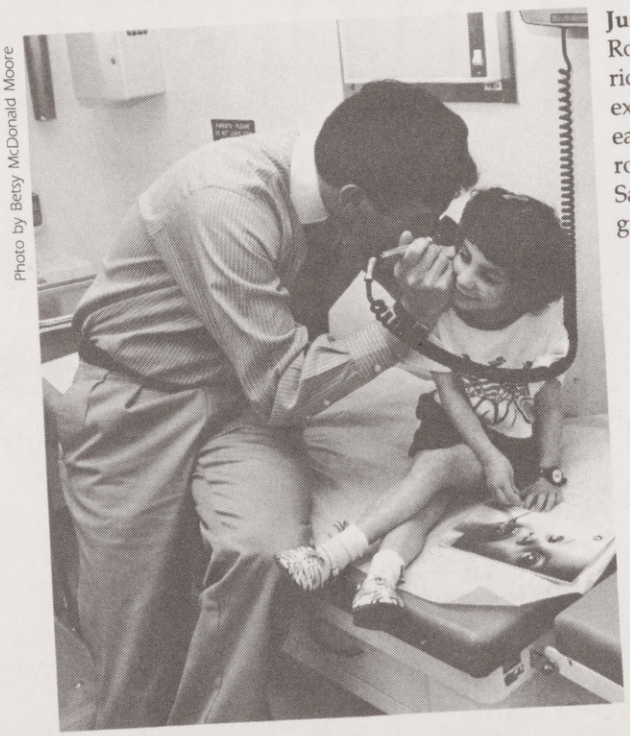
estimated that 10,000 to 15,000 children in the United States suffer from growth hormone deficiency.

Growth hormone is a protein that is produced by the pituitary gland. The pituitary gland is located in the middle of the brain and is connected to a portion of the brain called the hypothalamus.

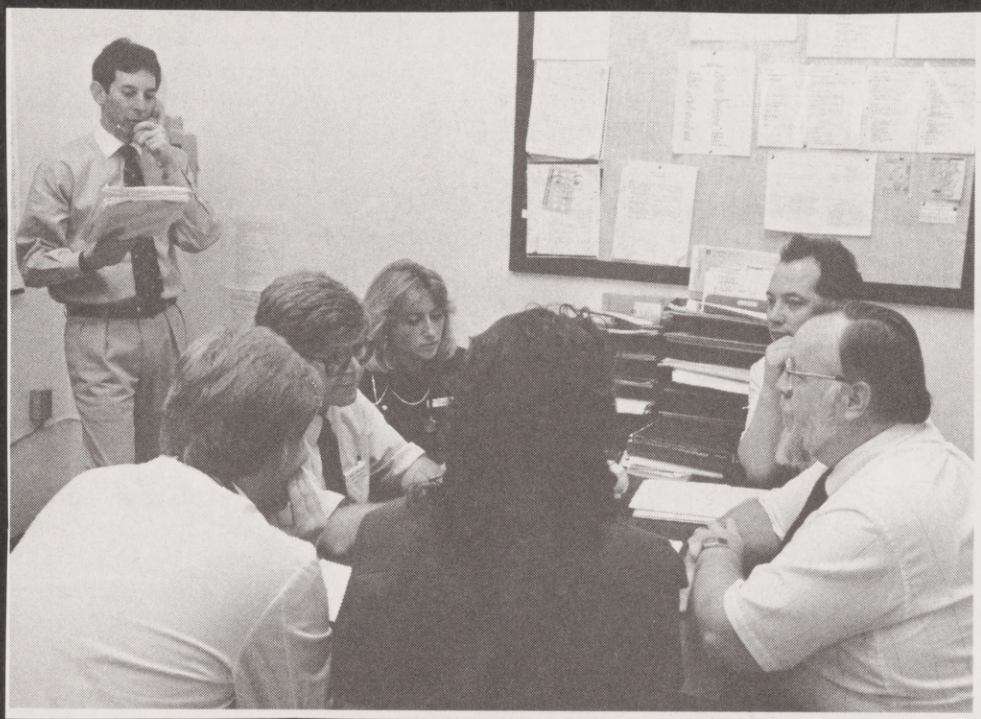
Approximately 2 million children in the United States are very short, with heights below the third percentile for their age. Many have serious but treatable growth disorders.

The history of using human growth hormone goes back 30 years according to Raymond Hintz, MD, pediatric endocrinologist at Chil-

Photo by Betsy McDonald Moore



Just a peek: Ron Rosenfeld, MD, pediatric endocrinologist, examines Sarah Garcia's ears as part of her routine clinic checkup. Sarah suffers from a growth disorder.



Helping their patients grow: Members of the Division of Pediatric Endocrinology discuss patients' progress at weekly meetings in Clinic A. Seated (from left) are Wayne Sells, MD, pediatric intern; Darrell Wilson, MD; Laura Bachrach, MD; Edgar Molina, MD, visiting physician from Guatemala; Raymond Hintz, MD, division chief; Kathy Morrison, MD, fellow; and standing (at left), Ron Rosenfeld, MD.

dren's Hospital and professor of pediatrics at Stanford University School of Medicine.

In the late 50s, researchers discovered that human growth hormone was useful in treating children with short stature, particularly those with pituitary problems. But, unlike other hormones that can be extracted from animal organs, growth hormone had to be obtained from humans. Therefore, the supply was never adequate.

"Only half of the children who were severely affected were able to get treatment," Hintz says. "Those who got treatment sometimes did not receive it long enough, or did not receive as high a dosage as needed because the supply was so restricted."

About 10 years ago, scientists at Genentech, Inc., in South San Francisco successfully cloned the gene that codes for growth hormone. The gene was then spliced with the genes of E. coli bacteria which multiply rapidly. This made possible unlimited amounts of synthetic human growth hormone.

In April 1985, pituitary-derived growth hormone was removed from distribution in the United States after several patients who had been treated in the 60s and 70s died of a rare brain virus.

Fortunately, in October 1985, Genentech received FDA approval to market the first biosynthetic growth hormone. Today, about 10,000 children nationwide are being treated with synthetic growth hormone.

Hintz and his colleagues in the Division of Pediatric Endocrinology—Ron Rosenfeld, MD; Darrell Wilson, MD; and Laura Bachrach, MD— are involved in three studies focused on long-term use of the synthetic hormone.

The group continues to follow children who have been treated with

synthetic growth hormone over the past six to eight years.

"We've had the chance to be pioneers in the field of synthetic growth hormone," Rosenfeld says. "We were the first place to test it. Now we're exploring disorders other than classic growth hormone deficiency that might benefit from growth hormone therapy."

Rosenfeld is exploring the effects of synthetic growth hormone on children with Turner's syndrome, a genetic disorder marked by short stature. About 50,000 females in the United States have the disorder, which involves a missing or defective X chromosome. Those girls reach an average adult height of 4 feet 8 inches.

Initial results indicate that growth hormone therapy can be effective in treating Turner's syndrome.

Another study involves children who do not suffer from growth hormone deficiency but who are still severely short in stature.

"These children are very short and are relatively delayed in their physical development," Hintz explains. "They are at a physical and psychological disadvantage compared to their classmates.

"If it can be shown that growth hormone therapy is useful for this group, it might become accepted treatment for children who do not have a growth hormone deficiency but are still very short," Hintz observes.

No new patients are currently being enrolled in these studies.

Although the drug is relatively expensive—up to $15,000 per year—most families are covered by insurance that pays for the treatment.

Short stature must be diagnosed and treated early in childhood to be treated effectively.

"Once a child goes through puberty, the cartilage centers in the bones close, and they are no longer able to grow in height," Hintz explains.

"No matter how much growth hormone you take then, you won't grow taller. You might end up with a big nose, but that's about it."

Hintz urges parents to schedule regular pediatric checkups for their children. "Many pediatricians don't see these children until they are brought in for their kindergarten or first grade physical."

Sarah grows up with help from synthetic hormone

"I just want to be called Sarah—not 'Little Sarah,'" says the dark-haired, 7 1/2-year-old pixie.

Sarah Garcia of San Jose has always been small, but now she is catching up to her younger sister, Megan, and her classmates—who average 18 inches taller than herself—thanks to synthetic growth hormone treatments.

Although she was a full-term baby, Sarah weighed only 3 pounds, 8 ounces at birth.

"Failure to grow normally in the womb is called intrauterine growth retardation," explains Ron Rosenfeld, MD, Sarah's pediatric endocrinologist at Children's Hospital.

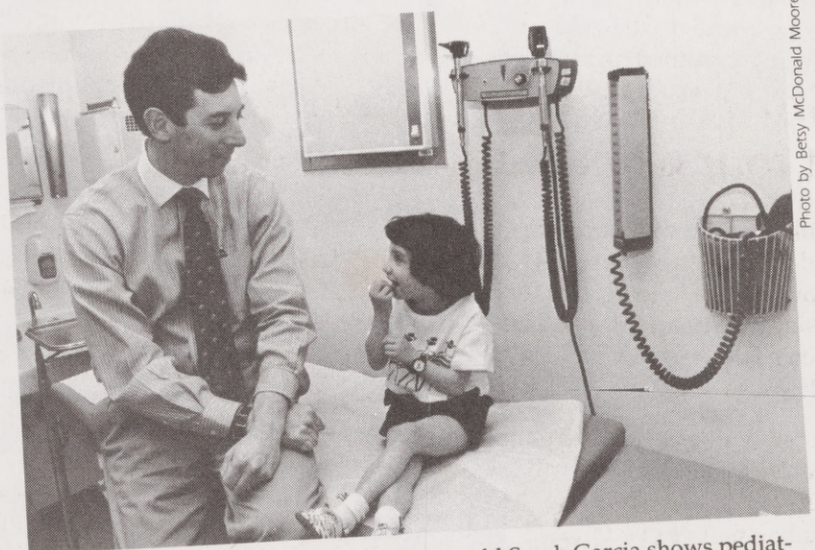


Photo by Betsy McDonald Moore

Playing grown up: Seven and a half year-old Sarah Garcia shows pediatric endocrinologist Ron Rosenfeld, MD, how "grown up" she is by putting on her play lipstick. Sarah is being treated with synthetic growth hormone for her severe short stature.

Sarah...Cont. from 4

"We still do not understand the cause of Sarah's short stature. She appears to produce enough growth hormone, but her bones don't respond normally."

Sarah's parents, Jose and Jane, have been bringing her to the Pediatric Endocrinology Clinic at Children's Hospital since she was 18 months old. She began treatment with synthetic growth hormone when she was four, soon after the drug received FDA approval. She has shown a partial response to the drug—growing approximately a half inch every six months.

Sarah herself prepares a syringe each evening and injects the hormone into her doll-sized upper thigh.

"It doesn't hurt, really," she assures. "I just get nervous if people watch me do it."

"Sarah fends for herself very well," her proud father, Jose, says. "Unlike some short children, she has a great self-image. She gets tremendous support from our big family as well as from the staff at Children's Hospital. They've been super."

Photo by Betsy McDonald Moore

Sarah Garcia

The family has made special accomodations for Sarah's short height. A stepping stool is kept at the bathroom sink for her to brush her teeth. Her dad created a special switch that enables her to turn the lights on and off. Until recently, she used a booster seat at the dinner table or when eating at a restaurant.

At school, Sarah says, someone has to hold the water faucet at the drinking fountain or at the sink while she stands on her tippy toes to get a drink or wash her hands.

Still, Sarah thinks that being short has its benefits.

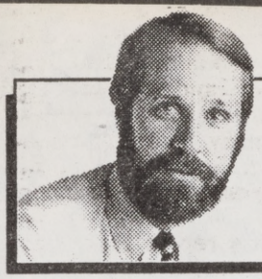
"She's extremely agile—climbs like a monkey," her dad comments.

"I love ballet and gymnastics," volunteers the 22-pound Sarah.

In July, the Garcia's plan to have a "Three-Feet Party" for Sarah. They estimate that she will reach the top of the yardstick by then.

Sarah will continue taking synthetic growth hormone as long as the drug continues to help her grow.

"People won't call me 'Little Sarah' anymore," she grins. "I'll get bigger and bigger 'til I get all the way up to my sister. Then they'll call me 'Big Sarah.'"



Pat Dillon

Handicapped by a definition

BEFORE US lies a social frontier, the lightly explored legal question of what exactly is a parking meter, anyway?

Is a parking meter the device that stands there, stationed at the curb, crudely ticking down a relatively puny amount of rental time, or does it record more, as a clock or calendar or sun dial would?

If Glenn Mulcihy of Cupertino doesn't get a straight answer, he's considering a class action suit on behalf of himself and other disabled motorists, who, he believes, are being denied their rights to park without penalty.

Mulcihy (that's Scottish and Irish, which don't blend smoothly — hence the strangely spelled last name and a somewhat feisty nature) is a 34-year-old child counselor who, back when he was a young chopper pilot, managed to get himself into the tail end of the Vietnam War just in time to get shot up.

He gets around on a couple of torn-up legs with the aid of a cane, and he carries a yellow disabled driver permit the DMV routinely issues.

"This is not a wounded vet issue," he said Tuesday. "What just happened to me could have happened to my grandmother, anybody."

What happened was this:

Mulcihy was late last Friday for a flight out of San Francisco International Airport to Milwaukee. As he entered the admittedly unfamiliar airport in his 1986 Corvette, he noticed first the sign for departing flights and then the sign that said "Parking."

"I LOOKED for the blue handicapped slots but they were full," he recalled. "So I started circling up and up, still looking for handicapped parking. There were no slots left. I went all the way to the top and parked in a regular zone right next to the elevator."

He returned five days later and when he reached the parking garage, he picked up a white courtesy phone to alert the toll takers that he was a disabled driver about to come to the gate.

More than an hour later, after angry debates with, he guesses, more than half a dozen airport officials, Mulcihy paid $78, the full parking fee, before he could leave the garage.

"No one would take responsibility for letting me through, even though my license clearly says I have the right to park for free," he said. "At one point, someone on the police chief's desk even sent me to the wrong terminal for someone else to pass the buck."

He read me the back of his license and cited DMV regulation 21458, which specifies where disabled drivers may park without penalty. This includes metered spaces. It does not say "at meters." It says "metered space."

"Doesn't matter whether you insert 50 cents into a slot or punch a card into a time clock or hand it to a toll taker. Three minutes, three hours or three days. It's all metered."

When I first brought this to the attention of Ron Wilson, the PR honcho at SFO, he sounded conciliatory, saying it was unfortunate that Mulcihy hadn't driven past the "Parking" sign and continued into the terminal area where handicapped parking is provided in outdoor concourses between terminals.

Then, he offered to refund the $78.

WHEN I called Mulcihy to relay the news, he reminded me that "money is not the issue. It is how disabled drivers are treated. And I was treated better by the lowest-paid people, the porters who helped me with my baggage cart, than I was by the people who set and enforce policy."

Out of curiosity, I posed the same circumstance to Jill O'Brien in the community affairs office at the San Jose airport. She said that, while there are disabled zones provided in the short-term parking area, a fee is charged, but it's at the cheaper, long-term rate.

Later Wednesday, I got back to Wilson at SFO. By then, he had requested and received an opinion from the San Francisco City Attorney's Office.

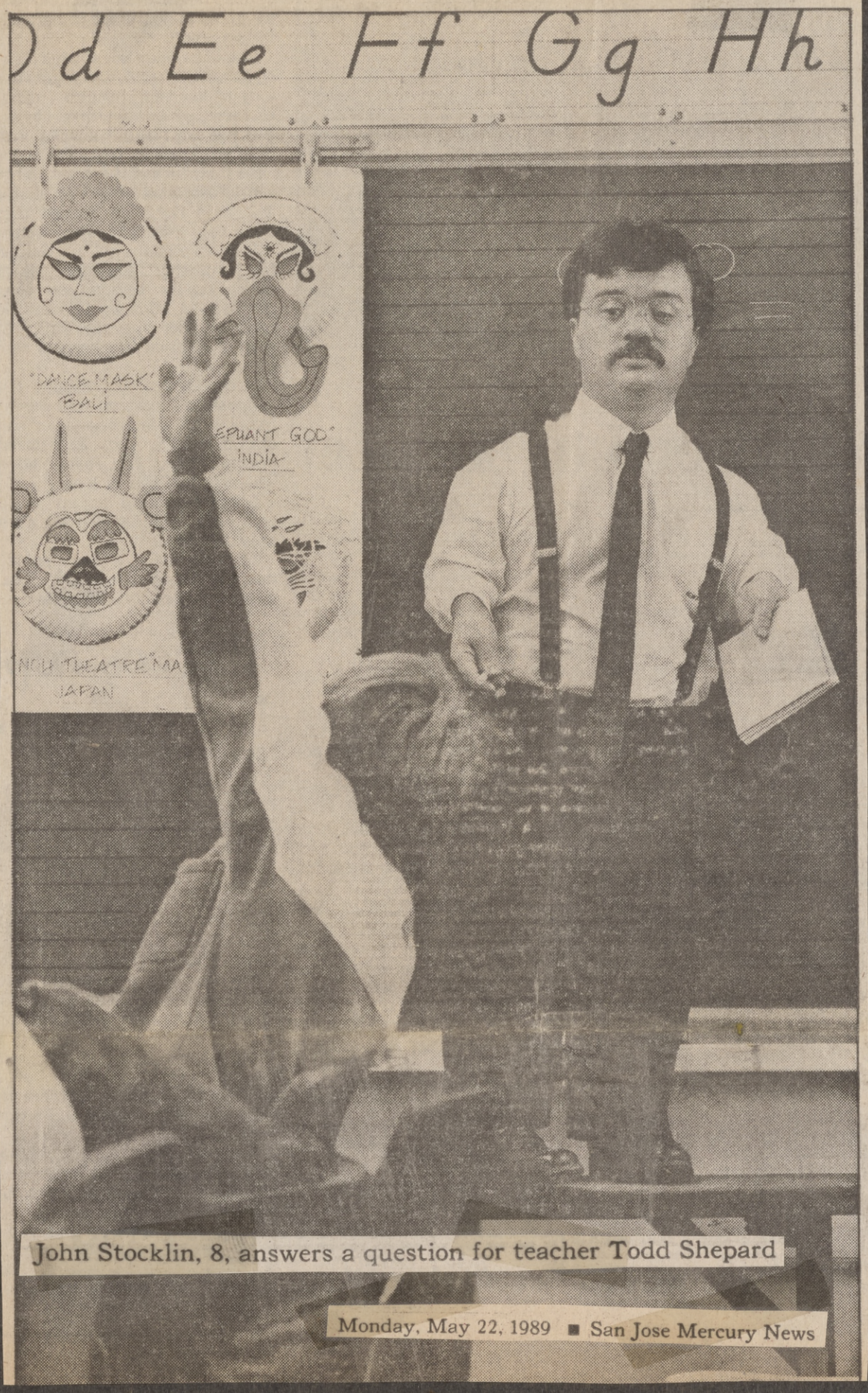
"The airport has designated parking for disabled drivers," he said, trying to sound friendly but firm. "The airport does not have metered zones. Thus, we are under no obligation to reimburse this person. I have been told that, under the circumstances, I was wrong to suggest that we would."

Earlier Wednesday morning, I had called the California DMV for a clearer definition of the term "metered space."

Bill Gengler of the public affairs office in Sacramento called me back to say that "under a strict interpretation of the law, where the time has a limit — that's a meter. Where there is no limit, it is not a meter."

I haven't called Glenn Mulcihy to report the official DMV version. But I know what he'll probably say. It's what you're probably saying right now.

Academic heights



John Stocklin, 8, answers a question for teacher Todd Shepard

4 feet 2 is no measure of P.A. teacher's stature

ENGROSSED — David Siddall, 8, talks with Todd Shepard about lesson on masks.

Todd Shepard's second-graders aren't being smug when they look down on him.

It's just that their teacher is 4 feet 2 — shorter than all but two of his 28 students at Palo Verde School in Palo Alto.

But they still look up to him for leadership. And as Shepard ends his first year as a teacher, the 31-year-old ex-computer worker has proven he has what it takes.

His students greet him with the singsong chant of "Good Morn-ing Mr. Shep-ard" that has probably greeted teachers since the Middle Ages. They look to him for approval when they've answered a question correctly and sneak forlorn glances his way when they've erred. He is, undoubtedly, in command.

Yet he can rarely escape being reminded of his tiny frame. He fits comfortably in a child's chair, making his mustache and deep voice seem incongruous.

For teacher, size not key to stature

TEACHER, from Page 1A

Sometimes ignorance and cruelty reign, such as when people point at him in supermarkets. Depending on who's doing the gawking and his mood, Shepard might explain he is a little person — he shuns "dwarf" as too clinical — or point back. "Or they'll say, 'Look, a midget!' " he said. "And I'll say 'Where?' "

The word midget offends him; he says it was coined by circus folk for little people who were clowns.

Rather than resort to bitterness, Shepard tries to show he is no different from anyone else. He also tries to pass on to his charges the self-esteem that many influential teachers and his parents, a Stanford University psychology professor and a lecturer in children's literature, instilled in him.

"The deal with self-esteem is mainly realizing that it's most important for people to realize that we all are different, but we all have positive things to contribute," Shepard said.

Praises innovation

What he has to offer shines as he helps a new reader sound out a word. Shepard questions students' comprehension as they wend their way through Weekly Reader and praises innovative answers.

> "It's just the way he is. It doesn't matter if he's small, or big or medium."
>
> *— Rachael Kleidon, pupil*

To many students, Shepard's gender is more of an issue than his height.

"I had to get used to Mr. Shepard because I hadn't had a boy teacher yet," Beth Aufdemberge said. "And I never had a little person who's been a teacher."

Added her pal Rachael Kleidon: "On the first day of school, it was fun, and it still is and our activities are very fun. I just treat him the same way I would another teacher. It's just the way he is. It doesn't matter if he's small, or big or medium."

SHOW AND TELL — Gosha Samotyj, 8, displays the mask made with the help of teacher Todd Shepard, who helps her as she explains it to the class.

Wanting to help others led Shepard into teaching. And by all accounts, teaching has been as rewarding for him as it has been for his students.

"I think it's been a real good year," said Barbara Smith, whose daughter Melinda is in Shepard's class. "No matter what, it's been a great experience for kids."

Shepard is not a pioneer in choosing teaching as his profession, said Harriet Stickney, spokeswoman for Little People of America, a national support group based in San Bruno.

Must establish discipline

As a little person, being a teacher "is a challenge," said Stickney, who taught for 12 years. "You have to establish discipline, because if your class is going to fall apart it might be blamed on the fact that you are a little person."

Discipline and setting the tone in class is especially difficult for first-year teachers, said Ruth Carleton, a first-grade teacher at Palo Verde.

"With many beginning teachers, it's very hard to get a handle on who you are," she said. "And you are the teacher. Getting the children to realize you are the person in charge is always very, very difficult. He has overcome that."

When Shepard started last fall, he "had some trouble with kids listening to him," Smith said. But when he realized the classroom regulations he had posted weren't effective, he had the class help him write new ones.

The revised rules are simple, calling for students to follow directions and not talk when someone else is speaking.

When the class becomes a little noisy, Shepard simply stops. He folds his arms across his chest and waits. It doesn't take long.

During a recent multiplication lesson, he commanded the class from his perch — standing atop a picnic bench. He also uses a bench when he wants to write high on the blackboard.

'Student of the Week' event

Implicit in Shepard's classroom is that each child is important. Each week the spotlight shines on a different "Student of the Week." The chosen student calls the others to recess, leads the line and runs errands to the office. Classmates draw pictures for (often of) the student, which are displayed on a central bulletin board.

Other details of classroom decor also are testament to self-esteem. Shepard had all of his students make posters depicting their good points. Eric Gray's had some dinosaurs and stars and this self-evaluation: "hard-working, neat, dependable."

> "We all have positive things to contribute."
>
> — *Todd Shepard*

If Tom Steege, principal of Palo Verde, drew such a poster about Shepard it would be equally flattering. Steege said he hired Shepard because "I consider him an intelligent person, capable of learning."

He should know. He remembers Shepard as a student at the former Stanford Elementary School, where Steege was principal. "He had enough courage within him that he could make his way," Steege said. "He stands up for himself."

Shepard said he was not teased much as he grew up on the Stanford campus. His parents expected him to be treated as everyone else, and Shepard was never segregated in special classes.

"The only way that I felt like it got in my way was I was always picked last for sports," Shepard said, which hurt because he harbored dreams of making varsity teams.

Shepard still enjoys sports — especially basketball. He is a proficient shooter.

After graduating from Gunn High School, Shepard attended the University of California, Davis.

"I knew I wanted to work with kids, or help people in some way," Shepard said.

But when he graduated in 1981, there was a glut of teachers. So he became a computer programmer and worked for Lockheed for four years.

"I finally reached a point where enough was enough, and I enrolled in a credential program," he said.

Making that move also impressed Steege: "The change from computers showed that he had a mission and that he held children higher than the almighty dollar.

"I like to see a man on the staff. And it's a society of diversity, and children need to see that at an early age."

Being little, Shepard said, is "considered a handicap. The interesting thing about being a little person is that I don't feel handicapped per se. I feel handicapped to the extent that there's things I can't reach or whatever. But in the end that's not a big deal."

ASSOCIATED PRESS

Rep. Steny Hoyer, *D-Md., shakes hands with Janet Dorsey of Springfield, Va., after the House passed a bill guaranteeing equal access to employment, transportation and other rights for disabled Americans.*

DISABILITY BILL

Highlights of the Americans with Disabilities Act, approved by the House Tuesday:

Who's covered

The bill defines a person with a disability as an individual with a physical or mental impairment that substantially limits him or her "in some major life activity." That covers an estimated 43 million Americans, including those with AIDS and the virus that causes AIDS.

Key provisions

▸ Discrimination against the disabled in hiring and on the job would be prohibited. The requirement would be phased in over two to four years, depending on the size of the company. Companies with fewer than 15 employers would be exempt.

▸ Hotels, restaurants, shopping malls, drugstores and business and professional offices, along with any other "public accommodations," would have to be made accessible to disabled people.

▸ Transportation, public and private, would have to be accessible. Fixed-route bus systems would have to be accessible for people with wheelchairs. Amtrak, Greyhound and commuter rail systems are covered.

▸ Speech- and hearing-impaired people would have access to telephone service by means of relay services. These services would have to be provided by phone companies within three years.

What's next

The bill goes to a conference committee to resolve differences with a Senate version passed last year. The major difference is a House provision that prohibits people infected with AIDS from handling food, although the disease has not been shown to be spread by food handling.

San Jose Mercury News, Wednesday morning, May 30, 1990

A pill that sheds fat, builds muscle isn't as near as your drugstore — yet

PHYSICAL ALCHEMY

By Donna Alvarado
Mercury News Staff Writer

A DRUG that turns soft, mushy bodies into lean and lanky physiques sounds like the ultimate fitness fix.

Bodybuilders and athletes swear they've been enhancing their bodies for years with illicit supplies of a substance called human growth hormone. Now scientists are cautiously exploring the legitimate side of this powerful hormone's potential to sculpt the human body.

In certain people, growth hormone appears to whittle fat and build muscle. The effects, so far, have been only mild in overweight dieters but remarkable in soft-bodied people who suffer from abnormally low natural levels of growth hormone.

"Growth hormone establishes the principle that we can remodel the human body," says Dr. David Clemmons, an endocrinologist who has studied the hormone's effect on hugely overweight dieters. "I think we're headed down the right road, but we don't have the right compound yet."

Clemmons says he thinks researchers will have to tinker with growth hormone before it can help people who are overweight. "We think we've given it every way but Sunday, and we haven't found enough acceleration of fat loss to make it worthwhile for dieters," he says.

But one professional bodybuilder says athletes have been building muscles and shedding fat for years now with illicitly obtained doses of the drug. Growth hormone is chemically different from steroids — another kind of muscle-developing drug abused by some athletes — and the choice of the elite who can afford to pay several thousand dollars for a three-month supply.

"They can do all the research they want on it," says the bodybuilder, Cathey Palyo of Santa Rosa, of the hormone's sculpting ability. "We've known that for a while."

Palyo says she does not take human growth hormone because of its potentially drastic side effects. "But a lot of athletes are kamikazes. All they see is the gold medal."

No one doubts that it is a powerful substance. Human growth hormone, produced naturally in the brain's pituitary gland, is what makes children triple in height from infancy to adulthood. About five years ago, scientists learned how to make synthetic versions of the hormone.

Now it's used to treat abnormally short children. Meanwhile, researchers are studying other powerful uses for this drug — which can cost $10,000 for a year's prescription from Genentech of South San Francisco, one of the two biotechnology companies licensed to market it for children.

Clemmons and other researchers say there is no doubt that the hormone does nudge the body to shed fat and retain muscle in certain circumstances. Although the effect in dieters is small, it is dramatic in normal-weight adults who don't make enough natural hormone themselves.

"We've proven the effect," he says.

Growth hormone could promise miracles for dieters one day

Help for the elderly?

One of the most tantalizing questions is what growth hormone might do in the elderly. As people age, they make less and less growth hormone, and grow more mush-bodied and brittle-boned.

"Many people have a growth-hormone menopause after the age of 40," says Dr. Daniel Rudman, a geriatric specialist at the Medical College of Wisconsin. "By age 40 or 50, they virtually stop making growth hormone. It's a natural question to ask whether restoring them to a youthful growth-hormone status would reverse any of the events that occur during aging."

Scientists say they draw a distinct line between giving the hormone to people who have a medical need for it and healthy people who take illicit versions of it to boost athletic prowess. Doctors still don't know the long-term consequences of taking human growth hormone but do know some short-term dangers in people who take too much.

"This is all fascinating," says Dr. Fran Kaiser, a specialist in geriatric medicine at St. Louis University who is studying growth hormone in the aged. "But this is something that needs careful observation. This is not an accepted therapy."

In the wrong doses and in the wrong people, she says, human growth hormone can cause serious side effects. It can promote enlarged hearts, spleens or livers, encourage diabetes and cause a condition called acromegaly that elongates the tips of certain bones.

"What people are studying is whether some aspects of aging . . . have anything to do with the diminished amount of growth hormone," says Perlman, the Genentech research director. If it does, he says, the implications are tremendous for the large group of aging baby boomers: "Aging is going to be a major issue in the United States," he says.

Body changes

Rudman says he is looking at whether growth hormone can influence body composition, kidney function and bone density in the aged. So far, he has seen no side effects in people taking moderate doses.

But some say a drug to prevent aging sounds unlikely. "It just seems unrealistic to me that we can all stay young by taking growth hormone," says Dr. Louis Underwood, a pediatric endocrinologist who works with Clemmons.

It will probably be years before doctors know the repercussions of giving growth hormone to youngsters, old people or dieters over a long period.

In the meantime, most researchers criticize healthy athletes who obtain growth hormone on the black market — a practice illegal in competitive athletics — saying they are tinkering with a risky proposition.

"You need to be careful about pushing growth hormone for building muscle mass," says Kaiser, the St. Louis University researcher.

Miraculous effects

When used cautiously, it can work miracles. The drug clearly turns abnormally short children into average-sized ones. Because it also turns them from chubby cherubs into lean and lanky kids, researchers have begun studying its effect on adults.

Growth hormone is generally given by injection. So far, it appears to help some people and not others:

■ Adults who are deficient in natural growth hormone appear to gain muscle and lose fat when given synthetic versions of the hormone. One study of 24 adults in London, who lacked the hormone because of treatment for pituitary tumors, showed that they lost an average of 12 pounds of fat and gained about 12 pounds of lean muscle. They show no net weight loss.

■ Hugely overweight dieters, given synthetic growth hormone in addition to their own normal levels, show a very mild tendency to accelerate fat loss while retaining lean muscle. But researchers conducting this study say the effect was so slight that it does not appear useful.

"You might have to take $10,000 worth of growth hormone to lose an extra four or five pounds of fat," says Clemmons, an endocrinologist at the University of North Carolina. To get a bigger effect, he says, "You're going to have to get into an unacceptable dose that causes side effects."

■ Highly trained athletes may add muscle and shed fat when given the hormone, but scientists are divided on this question. One study done at the University of New Mexico found this effect, but other researchers are skeptical.

"This is one study out of many," says Dr. Andrew Perlman, a clinical director of research at Genentech, which manufacturers a synthetic version of the hormone. "There are other studies going on elsewhere that show no effect in exercisers. The answer is clearly not in."

■ Older people, who make less and less growth hormone as they age, are a group that logically could be helped. Several different studies are ongoing around the country, but results are not yet known.

NORTH IDAHO NEWS NETWORK Tuesday, July 24, 1990

North Idaho

Socializing is highlight for Little People

By SUZANNE O'GORMAN
Staff writer

Sure, there are the medical clinics, the board meetings, the athletic competitions, the workshops, but best of all...there are the nightly dances.

Sarah Davies, 17, was smiling broadly as she sat on a bench on the boardwalk with her new friend Sandy Flerchinger, 19, waiting to board a cruise boat for a group picnic at Huckleberry Beach.

The two girls chatted like old friends, and yet they are recent acquaintances, brought together by an organization committed to cultivating the potential they can achieve as adults.

Teens comprise a large percentage of the 550 "Little People" gathered this week in Coeur d'Alene for their annual national conference, and to Davies, they are all potential friends.

Attending her second convention, Davies is disappointed that some of the new friends she made at last year's, in Baltimore, didn't make it here in 1990. But there are phone calls and letters, and after all, this was only her first opportunity to participate in a "reunion," she said.

> "It's so sweet when average-size guys get down on their knees to dance with you, but it's so much fun dancing with someone your own height."
>
> —Sarah Davies

Davies hopes to attend every year now, and she points out that with each time, the annual reunion will include the chance to see more and more of her friends as each convention further expands her social horizons.

Not that Davies is short on friends at home in Vancouver, British Columbia. She is pretty, smart and vivacious, with an engaging smile and an easy laugh. But life is different among friends of average height, she admits.

"It's sweet when average-size guys get down on their knees to dance with you, but it's so much fun dancing with someone your own height," Davies said.

Her new friends agreed.

Flerchinger was nervous about attending her first convention...not knowing anyone her own age who would be here and not knowing what to expect.

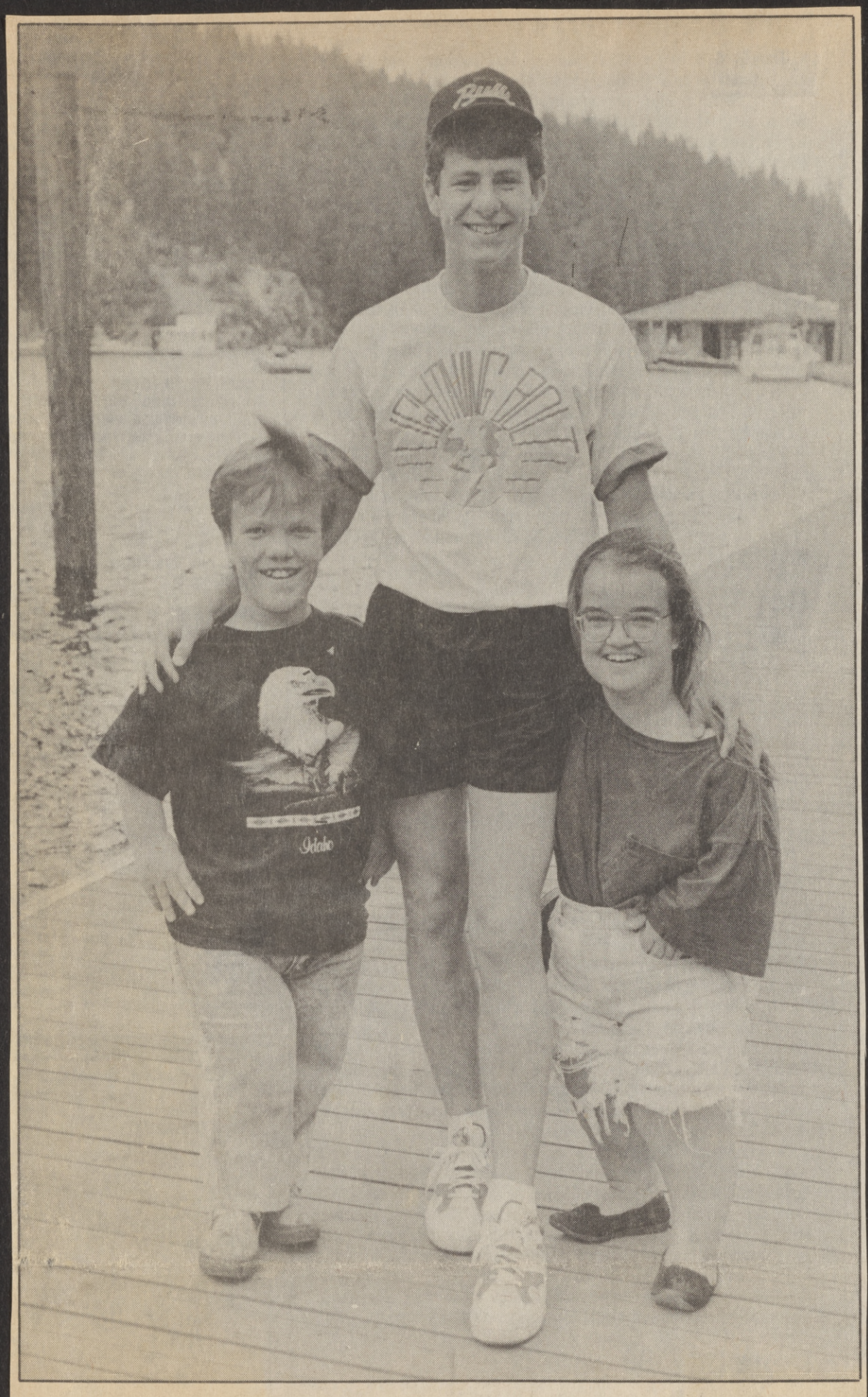
"But I started meeting people right away, and the more you get under your belt, the easier it is. I'm glad I came," she said.

Their advice to other teens and young adults of small stature when the next convention rolls around:

"Go for it," they advised. "The dances are the best."

Also giving high marks to the dances, convention first-timers Kelly Laing, 17, and his brother Johnny, 14, are having a "blast," they said. Johnny is the only little person among his family of 10, and his parents and three brothers, including Kelly, accompanied him to his first convention.

Attending as a family unit is encouraged for the obvious mutual support and shared enjoyment it provides, and as the younger Laing feigned pushing his brother off the pier, wondering aloud how deep the water might be, he was just a typical 14-year-old kid ... which is one of the points the Little People hope to spread to the taller in stature who may not understand.



—Press photo

Johnny Laing, 14, his brother Kelly, 17, both of Arco, Idaho, and their new friend Sarah Davies, 17, of Vancouver, B.C., are enjoying every moment of the national convention's social agenda.

Activist Richard Crandall calls dwarfism a disease of denial.

SHORT STORIES

Questions are seldom surprising

Orange County Register

Children and adults ask the same questions about dwarfs:

"Do you have little furniture?"

Yes. A company in Alabama makes adult-style furniture proportioned for a dwarf's body. Gracie Oliver and Richard Crandall have little chairs and a small, ornate love seat in the Huntington Beach house they bought, but most of their furniture is average-size.

"Do you drive a car?"

Yes, both drive cars. Oliver's is equipped with hand controls and a rack on the back to carry her wheelchair. Crandall drives a Dodge Caravan van. He uses pillows to support his back and for height. Blocks on the pedals enable him to reach the brake and gas.

"Do people knock you over because they don't see you?"

Yes. Oliver once was hit by a car backing up in a parking lot.

"How do dwarfs feel about dwarf-tossing?"

Dwarf-tossing is a contest in which little people wearing helmets are thrown by patrons of bars. It is insulting, demeaning and dangerous, Crandall said.

"You couldn't throw members of other minority groups and get away with it," Crandall said. "Why is it any different with us?"

Dwarf alters society's view of little people

By Laura-Lynne Powell
Orange County Register

HUNTINGTON BEACH — Richard Crandall couldn't believe he was putting himself through this ordeal. Again.

But he marched bravely into a Huntington Beach game room where young club members chased each other around a pool table. They stopped. A hush settled over the room. The kids stared.

A fourth-grader broke the silence.

"I'm taller than he is," the boy said, pushing out his chest and lifting his chin above Crandall's head.

Another, pointing to Crandali, announced: "He's a midget!"

Crandall hates that word. So do most dwarfs. It conjures up images of circuses and freak shows. They would rather be called short-statured, little or dwarfs, the medical term describing their stunted growth.

And though Crandall braces himself for taunts, he deliberately walks into situations where he might hear them. He lets the children, members of the Boys' and Girls' Club of Huntington Valley, stare in the hopes of ridding them of their fascination with his 3-foot-10-inch frame. Then he teaches a lesson in respect.

Since the 1983 suicide of his teen-age daughter, Crandall has dedicated his life to changing society's attitude toward little people. He blames dwarfism — and society's perception of it — for Michelle Crandall's struggle with alcoholism and her suicide. She left a note that read: "I guess I just couldn't handle the life God gave me."

After her death, he quit a successful sales career and founded the non-profit Short Stature Foundation in Irvine.

He visits schools, organizes sports events featuring a dwarf basketball team and lobbies to unite little people with the rest of society. His lesson to the children: He is little, but he is also a businessman and a parent, and he has plans to remarry.

The problems dwarfs face came to light last week after actor David Rappaport, 38, apparently killed himself in Los Angeles. In March, the 3-foot, 11-inch actor had been hospitalized after a failed suicide attempt. Though the actor's exact motives remain unknown, in a 1988 interview he complained about the discrimination and problems he had confronted because of his height.

On this day, Crandall ushered the children into a classroom and invited them to sit on the floor around him and his fianceé, Gracie Oliver, who also is short-statured. The couple sat comfortably in the small chairs built for kids.

"You can call me Miss Gracie and him Mr. Richard," Oliver said. "That is because we are adults. We are adults like your parents and your teachers."

The Huntington Beach couple told the children what it's like to be a dwarf — how hard it is to find clothes that fit; how frustrating it is not to be able to reach the spaghetti sauce at the market; how maddening it is to be laughed at by strangers.

When the lesson was over, a parent arriving early to pick up her son thanked the couple.

"I think what you're doing is great," she said.

Crandall smiled, clenched his fist and punched the air.

Success.

Richard Crandall's arms are disproportionately short, his fingers wide and stubby, his legs bowed. He brushes his wispy, graying hair away from his bearded face. His brown eyes glint when he smiles.

Crandall is 49, but he won't say so to strangers; it's a protest for always being asked. He likes to wear a worn straw cap, long-sleeve, tailor-made shirts and slacks held up by suspenders.

He was born in upstate New York in 1940 to average-size parents. The doctor knew right away Crandall would be different. His short legs and stubby fingers were a sign that the baby had cartilage-hair hypoplasia, a skeletal abnormality.

Of the hundreds of types of dwarfism, some are hereditary. Some dwarf couples, depending upon their type of dwarfism, have a 50 percent chance of having a dwarf baby.

But most dwarfism is caused by a random gene mutation. Ninety percent of dwarf babies are born to average-size parents.

Researchers say it is difficult to calculate the percentage of the population with dwarfism. Dr. David Rimoin, a leading geneticist specializing in dwarfism at Cedars Sinai Medical Center in Los Angeles, said the most common form of dwarfism, which affects 80 percent of disproportionate types of dwarfs, occurs in one in 10,000 births.

Crandall was the third of five children and the only dwarf. When he was 5, Crandall asked his father, "How come I'm not growing?"

He said, 'You're not going to grow, Richard; you're short.' That's it. That's all I was told."

Through most of his childhood, Crandall kept to himself. Bus rides home from school were agonizing because he was the brunt of other children's jokes.

Crandall learned how to make friends when he was in high school in Ontario, Calif.

He had been buying liquor since he was 13 because sales clerks couldn't figure out how old he was and were embarrassed to ask. He bought liquor for fellow students and became everyone's drinking buddy. His crude sense of humor made him a hit with the guys.

Girls, however, were another story. Crandall resented their rejection, so he made girls the target of his pranks.

"None of those girls were going to go out with me. So I hurt them before they could hurt me," he said.

Crandall calls dwarfism a disease of denial. He said he spent most of his life unable to accept his size and rarely socialized with other little people. That, he said, led to bitterness and a reliance on alcohol.

Crandall married one of the only dwarfs he ever dated in the early 1960s, and the couple moved to Orange County during their marriage. They had two children: Michelle, a dwarf, and Mykell, born two years later, who grew normally.

Crandall sold electronics for a living. By the late 1970s, he was making $200,000 a year. He invested in real estate in Laguna Beach, bought a 40-foot cabin cruiser and drove an expensive car.

But his professional and economic successes didn't stop people from staring. And that made Crandall mad.

Crandall said he became an alcoholic early in his marriage. The anger that had begun to well inside him was turned loose on his family.

His son, Mykell, who was taller than his parents by the time he was 10, remembers.

"My father was always belligerent," Mykell Crandall said. "He was mad at the world, and he wanted to get back at everybody."

Crandall's wife, Marilyn, filed for divorce in 1981.

His life began to change when he attended his first Alcoholics Anonymous meeting in May 1982. He had been sober for more than a year when his daughter committed suicide.

Michelle Crandall was having trouble making friends at Costa Mesa High School, where she was a junior. At 17, she was strawberry-blond with soft brown eyes and a cute, turned-up nose. But she was unhappy.

Crandall blamed society for her despair.

"There was no one to help us. There is still no one to help us," he said.

> "None of those girls were going to go out with me. So I hurt them before they could hurt me."
>
> **Richard Crandall**
> founder of Short Stature Foundation

After the suicide, Crandall decided it was time to show society the tragic consequences of its indifference.

In 1984, Crandall became a regional director for Little People of America, the largest support group for dwarfs in the world with 5,000 members. But soon he became disenchanted with the group.

He recently praised the organization's support for dwarfs but said it was ineffective at changing society's perception of little people.

In February 1984, Crandall opened his own non-profit organization for dwarfs, the Short Stature Foundation.

The Irvine-based foundation's goal is to reach dwarfs who aren't members of support groups. Fellowship, he believes, may help them avoid the years of alcoholism that plagued him and the anguish that caused his daughter's suicide. He quit his job to devote himself to dwarfs' problems, including:

- Lowering the 20 percent unemployment rate.
- Helping dwarfs hidden by their families adjust to independent living.
- Guiding alcoholic or drug-addicted little people toward counseling.

But he realized he couldn't do it alone. He enlisted the help of a popular, energetic woman he had met at a Little People party, Gracie Oliver.

Oliver, 33, has a crippling form of dwarfism that forces her to rely on crutches or a wheelchair. Standing, she barely reaches Crandall's shoulders.

Her loving style is captivating. It differs from that of Crandall.

"They'll be laughing and I'll get mad," Crandall said. "But she'll talk to them. She'll ask them if they like it when people make fun of them.

"It's like Custer talking with the Indians. She turns the whole situation around."

In the six years since opening the Short Stature Foundation, he has had successes and failures. He ran a pizza shop and a telemarketing agency that employed dwarfs. But pizza-making was too physically demanding, and telemarketing required more money than Crandall could raise.

Crandall ran the Short Stature Foundation without pay for three years with weekly bingo games helping to support the organization. Two years ago, he went on salary at $3,000 per month as executive director.

The foundation publishes a catalog of adaptive devices for dwarfs and disabled people — poles to reach light switches and utensils to remove jar lids for those with small or weak wrists and hands.

The catalog goes to 15,000 medical associations, dwarf groups and individuals.

Kira and Marty Brogden of San Diego understand the need for a pool of information on dwarfism. Soon after the birth of their son, Billy, now 5 months old, they learned he was a dwarf.

The couple was completely surprised, since there was no history of dwarfism in their family. They read everything they could find on the condition and were able to find expert medical help here.

> "The most important thing is finding other people with the same condition. I think it will help Billy growing up."
>
> **Kira Brogden**
> San Diego mother of dwarf

"And once we got over the initial shock, we told everyone we knew about it immediately," Kira Brogden said. "We wanted to educate them to understand that he is perfectly normal except for this. And we went to a meeting of Little People of America one week after he was diagnosed."

Now the couple is consideri moving to some place where life less hectic and their main concern finding medical expertise and su port groups elsewhere.

Crandall was able to supply the with material on this and Brogd said she is grateful.

"The most important thing is fi ing other people with the same c dition," she said. "I think it will h Billy growing up."

Crandall's major effort these da is to establish a nationwide toll-fr hot line offering information dwarfism.

With the support of several oth dwarf organizations, Crandall trying to raise $62,000 to pay for t hot line's first year of operati scheduled to begin July 15.

Sometimes the stress of findi funds and running the Short Statu Foundation gets to Crandall, and talks about ending his mission.

Friend Arturo Gil, a dwarf-act living in Alhambra, said he hop Crandall doesn't stop.

"He is trying so hard to make difference," he said. "I wish thin would go better for him. He's a sai He's the light at the end of the tunn for a lot of people."

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